Dermatology Practical & Conceptual

Pediatric Chondroid Syringoma: Systematic Review with High-Frequency Ultrasound Findings

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ABSTRACT

Introduction: Chondroid syringoma (CS) is a rare adnexal tumor, exceptionally uncommon in children. To date, only isolated case reports exist, and no systematic synthesis has been performed.

Objectives: We conducted a PRISMA-guided systematic review of pediatric CS and present an illustrative case that provides the first high-frequency ultrasound (HFUS) and elastography findings.

Methods: PubMed, Scopus, and Web of Science were searched through September 2025. Eligible reports included pediatric patients (<18 years) with histopathologically confirmed CS. Data on demographics, tumor location, diagnostic methods, treatment and outcomes were extracted. In parallel, an 8-year-old female with CS was evaluated clinically, dermoscopically, and with HFUS and elastography, followed by surgical excision and histopathology.

Results: The systematic review identified eight pediatric cases in the literature: three malignant and five benign. Most tumors arose in the head and neck region; two occurred in the extremities, including one in association with eccrine spiradenoma. All cases were managed surgically, with favorable outcomes in benign lesions, while malignant cases demonstrated recurrence, metastasis, and one death. Our case presented as a cheek nodule with dynamic dermoscopic changes and HFUS features of a round, lobulated, hypoechoic lesion with posterior enhancement, vascularity, increased stiffness, and a budding yeast–like appearance. Complete excision achieved an excellent cosmetic result without recurrence at one year.

Conclusions: This study provides the first systematic review of pediatric CS, consolidating current diagnostic and therapeutic knowledge. Our case adds the first detailed HFUS and elastography findings, underscoring the role of imaging as a noninvasive adjunct. Complete excision with long-term surveillance remains the cornerstone of management.

Introduction

Chondroid syringoma (CS) is a rare cutaneous adnexal tumor, first described as a mixed tumor of the skin by Billroth in 1859 and later designated as chondroid syringoma by Hirsch and Helwig in 1961 [1], and accounts for <0.1% of primary cutaneous neoplasms [2]. CS is considered a cutaneous "mixed tumor" of sweat gland differentiation, thought to arise from eccrine or apocrine units with a biphasic epithelial–myoepithelial component embedded in chondromyxoid stroma [3,4]. Typically seen in adults, pediatric cases are exceedingly rare, with only eight described in the English-language literature. Diagnosis is often challenging because clinical features are nonspecific, and most cases are confirmed only after histopathology. Imaging findings have been only sporadically reported, and ultrasonographic features are almost entirely absent from the literature.

To address this gap, we conducted a PRISMA-guided systematic review to consolidate the scarce pediatric literature on CS and summarize current diagnostic and therapeutic approaches. In addition, we contribute the ninth pediatric case, which, for the first time, documents high-frequency ultrasound (HFUS) and elastography findings, thereby expanding the understanding of this tumor's diagnostic characteristics and therapeutic implications.

Materials and Methods

The systematic review was conducted in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. A comprehensive literature search was performed on the PubMed, Scopus, and Web of Science databases through 3 September 2025. The search strategy used the following terms, adapted for each database: ("chondroid syringoma" OR "cutaneous mixed tumor") AND (pediatric OR child OR infant OR newborn OR neonatal OR adolescent).

All retrieved records were imported into a reference manager, and duplicate entries were removed. Titles and abstracts were screened, followed by full-text review of potentially eligible articles. Inclusion criteria were case reports or case series involving patients younger than 18 years with histopathologically confirmed cutaneous chondroid syringoma (benign or malignant) and sufficient clinical, diagnostic, and outcome data. Exclusion criteria were non-English-language studies where data could not be extracted, review articles without original cases, adult patients, tumors of salivary gland origin, lesions with alternative histopathological diagnoses, and cases with incomplete patient information.

To maximize capture, backward citation searching of included studies was performed using the Citationchaser web application, which also facilitated automatic duplicate removal. Newly identified studies were evaluated against the same inclusion and exclusion criteria. The PRISMA flow diagram illustrating the study selection process is shown in Figure 1.

For each included case, the following parameters were extracted: patient age and sex, tumor location, lesion size, clinical presentation, diagnostic modalities, histopathological and immunohistochemical findings, benign versus

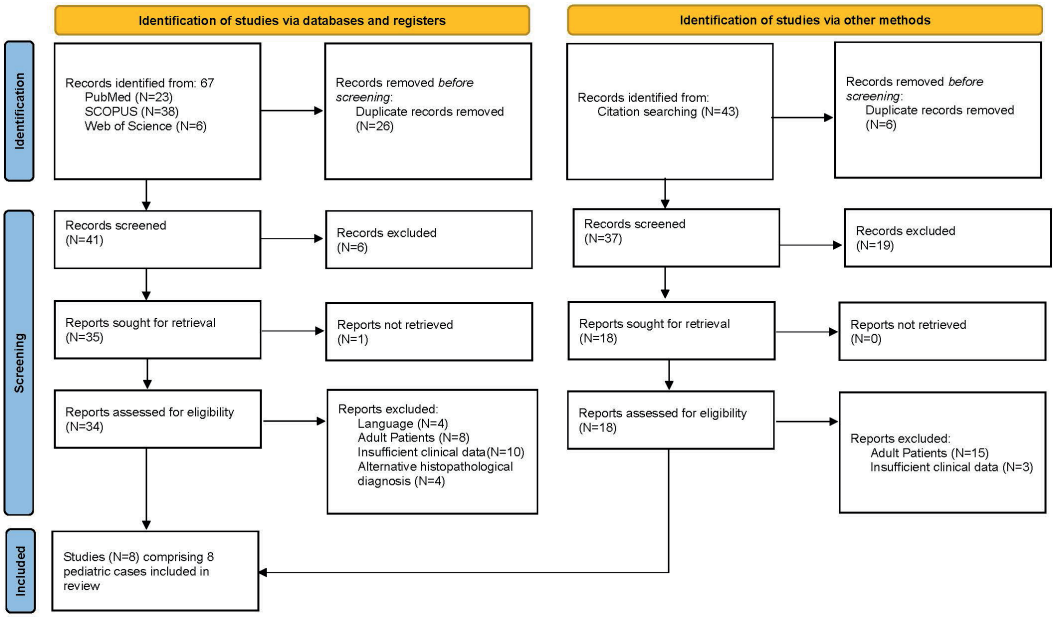

Figure 1. PRISMA 2020 flow diagram summarizing the study selection process. A total of 67 records were identified through database searching and 43 through citation chasing. After removal of duplicates and screening for eligibility, eight studies reporting pediatric patients with histopathologically confirmed cutaneous chondroid syringoma were included in the final systematic review.

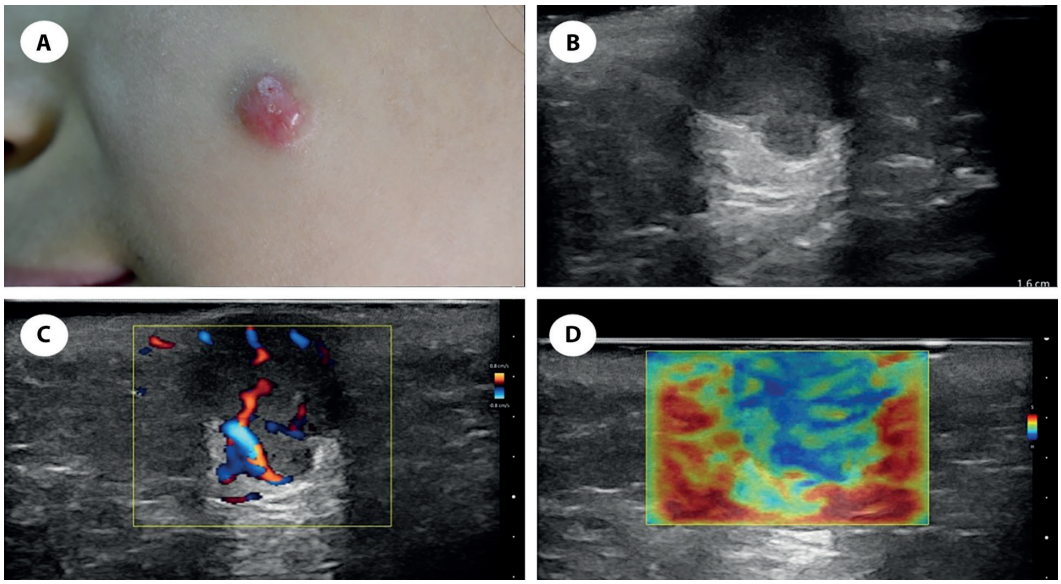

Figure 2. Clinical and ultrasonographic findings of the lesion. (A) Dermatological examination showing a well-demarcated, erythematous papular lesion on the left malar region. (B) High-frequency ultrasound demonstrating a well-defined, round lesion extending from the dermis into the subcutaneous fat, with posterior acoustic enhancement and a heterogeneous hypoechoic internal structure. A lobule protruding posteriorly into the subcutaneous tissue is observed (budding yeast-like appearance). (C) Color Doppler ultrasound showing central and peripheral vascularization. (D) Elastography map indicating increased stiffness of the lesion compared with adjacent tissue.

malignant character, treatment approach, recurrence status, presence of metastasis, follow-up and cosmetic outcome.

Case

An 8-year-old female presented to our clinic with a gradually enlarging mass on the left cheek. The lesion, which initially appeared as a small papule, had enlarged rapidly over the preceding four months. The patient reported no associated pain. Her personal and family medical histories were unremarkable, and systemic examination revealed no abnormality. Routine laboratory investigations were within normal limits.

Dermatological examination revealed a well-demarcated, erythematous papular lesion in the left malar region, measuring 6 mm in diameter (Figure 2A). The lesion was firm, immobile, and non-tender on palpation, with visible radial vessels on its surface. Dermoscopy showed a central milia-like cyst with radial vessels extending toward the center against an erythematous background. Diascopy produced an orange hue.

High-frequency ultrasound was performed in our outpatient clinic using a portable 20-MHz linear probe (Clarius Mobile Health, L20 HD3 Vancouver, Canada), including color Doppler and elastography. Imaging demonstrated a well-defined, round lesion measuring 7.7 × 6.9 mm, extending from the dermis into the subcutaneous fat. The lesion exhibited posterior acoustic enhancement and a heterogeneous, predominantly hypoechoic internal structure. A 2.6 × 2.0 mm lobule was also observed posteriorly, protruding into the subcutaneous tissue (Figure 2B). Color Doppler ultrasound revealed both central and peripheral vascularization (Figure 2C). Elastography demonstrated a firm lesion with increased stiffness compared with surrounding tissue (Figure 2D).

The patient's family expressed a preference for a watch-and-wait approach. Given the lesion's small size, benign clinical appearance, and the importance of minimizing invasive procedures and potential scarring in pediatric patients, we agreed on close monitoring. The patient was scheduled for follow-up after one month. At re-evaluation, the lesion had increased in size to 10 mm. Dermoscopy at that time demonstrated progression, with disappearance of the central milia-like cyst, development of a hyperkeratotic scale, and thickened radial vessels. These dynamic changes warranted further evaluation, and a punch biopsy was performed.

Histopathological analysis revealed a chondroid stroma containing focal tubular and ductal structures arranged in nests (Figure 3A). No cellular atypia, mitotic activity, or necrosis was observed. Periodic acid–Schiff with Alcian blue (PAS-AB) staining highlighted mucin within the stromal matrix, supporting the biphasic architecture of the lesion (Figure 3B). Immunohistochemical analysis showed positivity for p63 (Figure 3C), consistent with basal/myoepithelial cell differentiation, and S100 (Figure 3D), supporting myoepithelial involvement. These findings confirmed the diagnosis of chondroid syringoma.

The lesion was treated with complete surgical excision, considered the gold standard to prevent recurrence. The patient was followed up at three-month intervals. At the

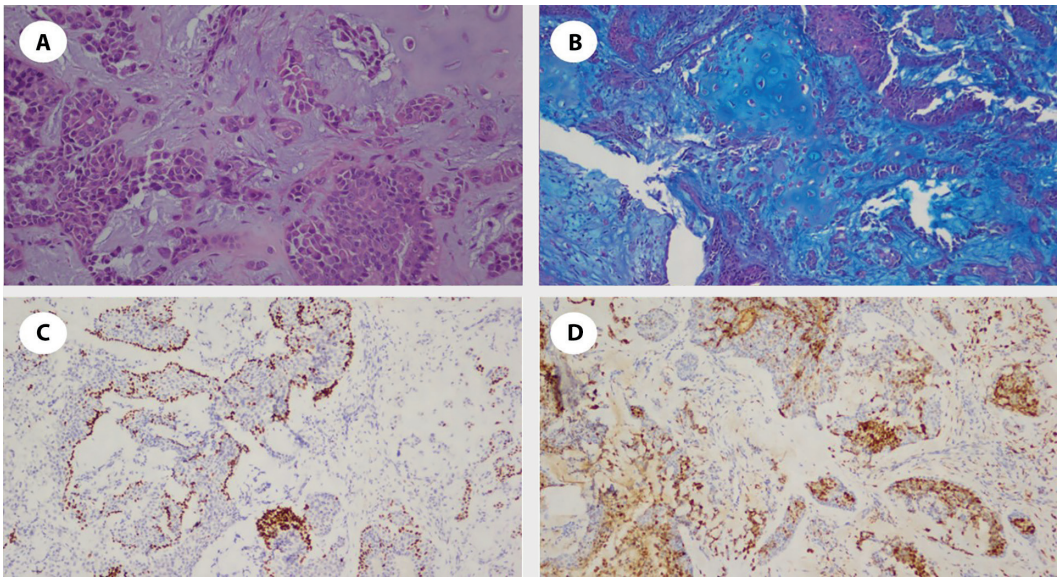

Figure 3. Histopathological and immunohistochemical findings of the lesion. (A) Hematoxylin-eosin staining showing a chondroid stroma with focal tubular and ductal structures arranged in nests (original magnification 10x). No cellular atypia, mitotic activity, or necrosis is observed. (B) Periodic acid-Schiff with Alcian blue (PAS-AB) staining highlighting mucin within the stromal matrix, confirming the biphasic architecture of the tumor. (C) Immunohistochemical staining positive for p63, consistent with basal/myoepithelial differentiation. (D) Immunohistochemical staining positive for S100, supporting myoepithelial involvement.

three-month follow-up, the surgical site showed only a minimal scar. At one year, there was no evidence of recurrence, and the cosmetic outcome remained excellent.

Results

The systematic review identified eight eligible studies [5-12] reporting pediatric cases of cutaneous chondroid syringoma. Together with the present case, a total of nine pediatric patients have been described to date (Table 1).

The patients' ages ranged from 7 to 17 years, with a mean age of 12.8 years. Five patients were male and four were female. Lesions were predominantly located in the head and neck region, including the cheek [11], nose [8,10,12], external auditory canal [6], and occipital scalp [7], although extremity [5,9] involvement was also documented.

Three of the nine cases demonstrated malignant histopathological behavior [5-7]. Two of these malignant cases developed both local recurrence and distant metastasis [5,7], and one patient died 34 months after treatment [7]. The remaining six cases were benign [8-12], with no recurrence reported during follow-up. Notably, one case described the coexistence of chondroid syringoma with eccrine spiradenoma in a Blaschkoid distribution of the lower limb [9], highlighting the potential for combined adnexal differentiation in pediatric patients.

Histopathology was the diagnostic standard in all cases. Imaging was rarely employed, with one report mentioning conventional ultrasound [9] without describing specific lesion characteristics, one utilizing magnetic resonance imaging (MRI) [11], and one including X-ray imaging [7] to assess metastasis. Our case is the first to incorporate high-frequency ultrasound with elastography, providing detailed characterization of lesion morphology, vascularity, and stiffness.

On high frequency ultrasound, the lesion appeared as a well-defined, round, lobulated nodule located at the dermal-hypodermal interface, with heterogeneous hypoechoic echogenicity and clear margins. Prominent posterior acoustic enhancement was observed. A small lobule was also noted posteriorly, protruding into the subcutaneous tissue and creating a budding yeast-like appearance. Color Doppler examination demonstrated both central and peripheral vascularization. Elastography revealed increased stiffness compared to surrounding tissue, consistent with a firm lesion. High-frequency ultrasound and elastography findings of the present case are summarized in Table 2.

Surgical excision was the treatment of choice in all cases. In one malignant case, excision was combined with radiotherapy and chemotherapy [7]. Follow-up periods ranged from six months to 17 years. Most patients remained alive with no evidence of recurrence [5,6,8,10,11]. Cosmetic outcomes were inconsistently reported; where described, they were generally favorable [8,11], and our case demonstrated an excellent cosmetic result at one year.

Discussion

Chondroid syringoma is a rare cutaneous adnexal tumor, with an incidence reported less than 0.1% [2]. It occurs

Table 1. Summary of reported pediatric cases of cutaneous chondroid syringoma, including the present case. The systematic review identified eight eligible studies describing pediatric patients with histopathologically-confirmed cutaneous chondroid syringoma. Together with the present case, a total of nine pediatric cases has been documented to date.

Case	Histopathological behavior	Age	Sex	Tumor location	Metastasis	Recurrence	Diagnostic modalities	Treatment	Follow-up and outcome	Cosmetic outcome
J. M. Hilton et al., 1973	Malignant	14	Female	Upper left arm	Yes	Yes	Histopathology	Excision	Alive with no recurrence (after 17 years)	Not reported
J. B. Botha et al., 1978	Malignant	15	Female	Left external auditory meatus	No	Yes	Histopathology	Excision	Alive with no recurrence (after 6 years)	Not reported
G. Hermann et al., 1987	Malignant	13	Female	Left occipital	Yes	Yes	Histopathology and X-ray for metastasis	Excision, local radiotherapy, chemotherapy	Deceased (after 34 months)	Not reported
N. Turhan-Haktanir et al., 2007	Benign	11	Male	Right lateral nose	No	No	Histopathology	Excision	Alive with no recurrence (after 10 months)	Excellent
A. K. Nath et al., 2009	Benign (Chondroid syringoma associated with eccrine spiradenoma)	14	Male	Right lower extremity	No	Not reported	Conventional ultrasound and histopathology	Excision	Follow-up not reported	Not reported
L. S. Solanki et al., 2011	Benign	16	Male	Nose	No	No	Histopathology	Excision	Alive with no recurrence (after 1 year)	Not reported
P. Purkayastha et al., 2021	Benign	7	Male	Left cheek	No	No	MRI and histopathology	Excision	Alive with no recurrence (after 6 months)	Excellent
S. Halder et al., 2024	Benign	17	Male	Tip of the nose	No	No	Histopathology	Excision	Follow-up not reported	Not reported
Present Case, 2025	Benign	8	Female	Left cheek	No	No	High-frequency ultrasound, elastography, and histopathology	Excision	Alive with no recurrence (after 1 year)	Excellent

Table 2. High-frequency ultrasound (HFUS) and elastography findings of pediatric chondroid syringoma. Summary of ultrasonographic characteristics reported in the present case, including lesion dimensions, echogenicity, vascularization, posterior acoustic features, and stiffness on elastography. HFUS demonstrated a well-defined, round, lobulated hypoechoic lesion with posterior acoustic enhancement, central and peripheral vascularization, and increased stiffness compared with surrounding tissue.

HFUS	Echogenicity	Location /Shape/ Margin	Posterior Acoustic Artefact	Doppler Vascularization	Elastography Findings
Chondroid Syringoma	Hypoechoic, heterogeneous	Dermal-hypodermal, round and lobulated (budding yeast-like appearance), well defined	Enhancement	Central and peripheral vascularization	Firm

most commonly in adults, whereas pediatric cases are exceedingly uncommon. To date, only nine pediatric cases, including the present report, have been documented in the English-language literature. To ensure comprehensive coverage, we performed a PRISMA-guided systematic review, which identified eight eligible cases in addition to the present one. The true incidence is likely underestimated as these tumors often present with nonspecific clinical features that resemble more common cutaneous nodules. Chondroid syringomas typically arise in the head and neck region [2], although extremity localization has also been described. Among the nine pediatric cases identified in our review, three demonstrated malignant histopathological behavior [5-7]; two located in the head and neck region [6,7] and one in an extremity [5]. Of the six benign cases, five involved the head and neck [8,10-12] and one was located on the right lower extremity in a Blaschkoid distribution, reported in association with an eccrine spiradenoma [9]. Taken together, these findings support the literature indicating a predilection for the head and neck region, while also highlighting that both benign and malignant tumors rarely present in the extremities.

The diagnosis of chondroid syringoma in pediatric patients is particularly challenging due to its nonspecific clinical presentation, which can mimic more common lesions such as pilomatricoma. Chondroid syringomas typically present as firm, asymptomatic, skin-colored nodules that are slow growing, solitary, and located in the dermal or subcutaneous layers, usually measuring between 0.5 and 3 cm in diameter [13,14]. Because many adnexal tumors share overlapping clinical characteristics, pediatric nodules are frequently misdiagnosed, and definitive diagnosis is usually achieved only after surgical excision and histopathological evaluation. Importantly, our systematic review revealed that one third of pediatric cases exhibited malignant histopathological behavior, a proportion that appears higher than what has been reported in adult series, though the numbers are too limited to draw definitive conclusions.

In previously reported pediatric cases, imaging was either absent or minimally described. One case mentioned the use of conventional ultrasound [9] without detailing lesion characteristics, another utilized MRI [11], and one described X-ray findings [7] in the context of metastasis. Beyond the pediatric literature, sonographic descriptions of CS are also scarce. Whittle et al. reported a finger CS imaged with high-resolution ultrasound, described as a solid, well-defined hypoechoic subcutaneous mass adjacent to the tendon, highlighting that CS can appear as a nonspecific hypoechoic nodule on ultrasound and may enter the differential diagnosis of slow-growing soft tissue lesions.[15] In contrast, our case provides the first detailed account of high-frequency ultrasound and elastography findings in a pediatric chondroid syringoma, adding valuable noninvasive diagnostic information to the literature.

On HFUS, the chondroid syringoma in our patient appeared as a well-defined, round, lobulated hypoechoic lesion with posterior acoustic enhancement, central and peripheral vascularization, and increased stiffness on elastography. A posterior lobule protruding into the subcutaneous tissue produced a budding yeast–like appearance, which, to our knowledge, has not been previously described. These findings can aid in distinguishing chondroid syringoma from other nodular lesions in children. Pilomatricomas, for example, often demonstrate a target-like appearance with a hyperechoic center, calcific foci, and posterior acoustic shadowing [16-19]. Trichilemmal cysts may contain hyperechoic linear structures corresponding to hair shaft fragments or keratin layers [17], while epidermal cysts sometimes exhibit a subepidermal connecting tract and can present with a "pseudotestes" or "onion-layer" pattern [16,20]. Hidradenomas typically show prominent hypoechoic solid and anechoic fluid-filled lacunar areas, and eccrine variants may be lobulated [21]. Dermatofibromas, in contrast, are ill-defined hypoechoic dermal structures that may extend into the hypodermis, but they lack lobulation and do not display convex

deep hypodermal margins [17,22]. Recognition of these comparative features may help clinicians include chondroid syringoma in the differential diagnosis when encountering pediatric nodular lesions.

Dermoscopy has been less frequently reported but generally reveals milia-like cysts, white streaks, telangiectatic vessels, and homogeneous erythematous backgrounds, sometimes with crown-like vascular arrangements [3]. Additional features such as brown blotches or a marble-like white-red pattern have been described [3]. In our case, dermoscopy initially showed a central milia-like cyst with crown-like radial vessels over an erythematous background. At one-month follow-up, the cyst disappeared, accompanied by a hyperkeratotic scale and thickened vessels. Such dynamic changes in dermoscopic appearance have not been emphasized previously and may serve as a useful clinical clue.

In pediatric facial nodules, dermoscopic differential diagnosis is broad and should include Spitz nevus, pilomatricoma, juvenile xanthogranuloma (JXG), pyogenic granuloma, and idiopathic facial aseptic granuloma (IFAG). Spitz nevi may show a starburst pattern in pigmented lesions or dotted/glomerular vessels with white lines in amelanotic variants, features not observed in our case [23]. Pilomatricoma classically demonstrates white or yellow-white structureless areas, often with blue-gray areas and characteristic vascular patterns; importantly, calcification-related clues on dermoscopy and ultrasound (e.g., echogenic foci with shadowing) help differentiate pilomatricoma from CS [18,24]. JXG frequently exhibits a yellow-orange background ("setting-sun" appearance) with linear vessels, which differs from the milia-like cyst and crown-like radial vessels seen initially in our lesion [25]. Pyogenic granuloma typically shows a homogeneous red structureless area with a white collarette and may display intersecting white lines ("rail lines"), aiding distinguishing it from adnexal tumors [26,27]. IFAG is a key clinical mimic on the pediatric cheek; dermoscopically, IFAG has been reported to show characteristic vascular and background features, and cutaneous ultrasound can further support the diagnosis by demonstrating a distinctive inflammatory nodule pattern, helping to avoid invasive procedures when clinical suspicion is high [28,29].

Despite their utility, dermoscopy and HFUS are not universally available in pediatric dermatology practice and have limitations. Both techniques are operator-dependent and require training and experience for acquisition and interpretation; availability varies across centers, and device cost may be a barrier, particularly for high-frequency probes and elastography-capable platforms. HFUS interpretation is also influenced by lesion depth, patient cooperation, and technical parameters, while elastography may show variability across devices and protocols, and standardized diagnostic thresholds for adnexal tumors are not established. Therefore, imaging and dermoscopy should be viewed as complementary tools that refine the differential diagnosis and guide management, but histopathology remains the diagnostic gold standard when growth, atypical features, or uncertainty persists.

Histopathologically, chondroid syringoma is defined by its biphasic architecture, combining epithelial and mesenchymal elements. Hirsch and Helwig described classic features including nests of cuboidal cells, tubuloalveolar and ductal structures, occasional keratinous cysts, and a variably composed stroma that may be chondroid, myxoid, fibrous, hyaline, or osseous [1]. Two variants have been recognized: an eccrine type with small, uniform lumina within a myxoid-chondroid stroma and an apocrine type with irregular branching tubular structures [4]. Immunohistochemistry typically demonstrates epithelial markers such as keratin and epithelial membrane antigen (EMA), while myoepithelial components often express S100, p63, and smooth muscle actin (SMA) [30]. In our case, histopathology revealed ductal and tubular structures within a chondroid stroma, with PAS–Alcian blue positivity confirming the biphasic nature. Immunohistochemistry showed p63 and S100 expression, consistent with myoepithelial differentiation, aligning with prior reports.

Total surgical excision has consistently been reported as the treatment of choice in pediatric chondroid syringoma, irrespective of histological subtype [5-12]. Benign cases generally remain recurrence-free, whereas malignant tumors carry a risk of recurrence, metastasis, and even death, as illustrated by one fatal case 34 months after treatment despite adjuvant radiotherapy and chemotherapy [7]. Our case was managed by complete excision with excellent cosmetic healing and no recurrence at one year. Cosmetic outcomes were inconsistently documented in the literature but were favorable when reported [8,11]. These observations support surgical excision as the gold standard and highlight the importance of long-term follow-up, particularly in malignant cases.

This review provides the first systematic synthesis of pediatric chondroid syringoma and outlines current diagnostic and therapeutic approaches. Most cases involved the head and neck [6-8,10-12], although rare extremity presentations were also reported [5,9]. Three of nine cases demonstrated malignant histopathology [5-7]; however, this proportion is likely affected by reporting bias. Our case also contributes the first detailed high-frequency ultrasound and elastography findings, expanding current knowledge of the imaging features of this tumor. Together, these observations enhance clinical awareness and support recognition, complete excision, and long-term surveillance as key elements in management.

Conclusion

Pediatric chondroid syringoma is an exceptionally rare adnexal tumor, with only nine cases, including the present report, documented in the English-language literature. By conducting a systematic review, we provide the first synthesis of pediatric cases, outlining their clinical features and diagnostic and therapeutic approaches, while also emphasizing the need for caution in interpreting malignant potential. The predominance of malignant cases reported is likely influenced by reporting bias, with malignant cases more often published than benign ones. Accordingly, the true risk of malignancy in pediatric patients remains uncertain, and additional reports will be necessary to clarify this issue. Our case additionally contributes the first detailed description of high-frequency ultrasound and elastography findings, thereby expanding the diagnostic framework for pediatric cutaneous nodules. Recognition of these imaging characteristics may facilitate earlier consideration of chondroid syringoma in the differential diagnosis, while complete surgical excision remains the definitive treatment. Long-term surveillance should be regarded as essential, as the biological behavior of this tumor in children remains incompletely defined.

Statement of Ethics and Patient Consent: The Declaration of Helsinki was followed during the investigation. The patient's parent gave written informed consent for the case report to be published, allowing for the release of the patient's medical records and any related photos. Upon request, a copy of the permission form is provided.

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