Chondroid syringoma: an unusual presentation in a 7-year-old boy

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SUMMARY
Coined in 1961 by Hirsch and Helwig, the term chondroid syringoma refers to a rare mixed tumour of subcutaneous tissue. Histologically, these tumours are almost identical to pleomorphic adenomas, arising from salivary glands. With the obvious difference being the presence of sweat gland tissue (syringoma) within a matrix of cartilage (chondroid). These mixed tumours remain scarce throughout the world, with an incidence of less than 0.098%. The vast majority of cases are reported in middle-aged and older adults, where they typically present as painless swellings in the head and neck, which gradually grow in size.

BACKGROUND
This case is unique because not only was the chondroid syringoma (CS) found in a 7-year-old child, but it presented as a rapidly growing swelling which tripled in size over a 4-month time period. We hope that by presenting this atypical case, CS remains a differential diagnosis for patients of all age groups, who present with a painless swelling.

CASE PRESENTATION
A 7-year-old boy was referred from his community paediatric services with a rapidly growing lesion that had tripled in size over a short duration of 4 months, and interfered with him wearing his glasses. The patient was urgently reviewed by the paediatric plastic surgery team and subsequently discussed with paediatric oncology, with the concern that the lesion may represent a malignancy. On examination, a 2×2 cm firm lesion was found in the left cheek. It had a blue discolouration with a surrounding area of hypervascularity. An MRI scan showed a well-defined mass, involving cutaneous and subcutaneous tissues, which were separate from the parotid gland and underlying musculature. An ultrasound-guided core biopsy enabled examination of the histological features required to make the diagnosis of a CS.

Under general anaesthetic, the left cheek was widely infiltrated with 0.25% bupivacaine+epinephrine (1:200 000). Via a circumferential incision, the lesion was excised with a 5 mm peripheral margin with a 5 mm cuff of subcutaneous fat. The skin was closed respecting the lines of election with 5–0 Monocryl (deep dermal) and a 5–0 vicryl rapide (continuous subcuticular). The excised specimen was 3×3×3 cm in size, and was sent for histological analysis, which confirmed the diagnosis of a CS. The lesion was well circumscribed with no infiltrative growth into surrounding tissues. There was no evidence of cellular atypia, increased mitoses or perineural invasion and no lymphovascular invasion was seen.

OUTCOME AND FOLLOW-UP
The surgical scar healed well and a follow-up ultrasound scan in 6 months displayed no evidence of tumour recurrence. The patient will attend clinic every 3 months for clinical review and also undergo ultrasound surveillance every 6 months.

DISCUSSION
CS is a benign neoplasm which is derived from sweat glands, and it typically presents as a painless subcutaneous nodule found in older and middle-aged
Case report

Figure 3  Histology slide confirming the presence of a chondroid syringoma.

Figure 4  A 6-month postoperative image following excision and reconstruction.

adults. With an annual incidence of less than 0.098%, CS remains a rare diagnosis which can only be confirmed after histological analysis. The condition is even less likely to be found in a child, with the first case not being reported until 2007. In their study, Turhan-Haktanir et al reported on an 11-year-old boy, who presented with a mass on his right nasal ala, which had been gradually growing for a year. Their presumptive clinical diagnosis was a pilomatrixoma (a benign appendageal tumour of hair follicles), and it was not until the excised specimen was sent for histopathological analysis, that they could come to the diagnosis of a CS.

Amin et al published a case of CS which bears some clinical resemblance to our case. They reported on a benign CS in the cheek of a 50-year-old woman, which had gradually started to interfere with her vision. The stark difference between the cases is that their patient’s CS tumour grew over a course of three decades, whereas our patient's tumour had tripled in size over a 3-month period to give any exposure to radiation. MRI shares this benefit at a higher cost, but does provide highly detailed images of the soft-tissues to aid diagnosis and surgical planning. Ultrasound-guided core biopsy was performed in this case on advice from the regional paediatric oncology unit and following the UK guidance on management of suspected primary soft-tissue sarcomas, in order to maximise diagnostic yield. It is worth noting however that fine-needle aspiration cytology (FNAC) can be used to establish the histological criteria required to make a diagnosis of CS. This is a less invasive method of analysis, which should be carried out before excision of the lesion. FNAC may be less appropriate in children, who will likely require some form of sedation but is particularly useful in the management of recurrent or metastatic malignancies of the head and neck.

Surgical excision remains the most effective method of treatment for CS, as it ensures the entire tumour is removed at once. In order to reduce the risk of tumour recurrence, it is also recommended to remove a rim of normal tissue along with the tumour. Less invasive methods have been reported, which include electrodesication and dermabradion, and even laser vapourisation. Despite the chosen method of CS treatment, it is usually recommended that patients are followed up regularly to detect any tumour recurrence.

CS typically presents as a benign tumour, but in a minority of cases it can take a malignant form, where the tumour is characterised by local invasion and more rapid growth. The most devastating cases of malignant CS have been shown to display widespread metastases, and their location is not limited to the head and neck, but can be found throughout the body.

Learning points

- In conclusion, we have presented an unusual case of chondroid syringoma found in the cheek of a 7-year-old boy.
- This case is notable not only due to the patient’s age, but due to the rapid growth of the tumour, which tripled in size over a 4-month time period.
- We hope that this case will raise awareness of the potentially varied nature of chondroid syringoma in patients of all age groups.
Malignant forms of CS will also have characteristic features on histopathological examination, which include a more pronounced presence of mitotic figures and tumour necrosis. In such cases, novel methods of comparative genomic hybridisation may be used to clarify the diagnosis. Some authors have even suggested that tumour surveillance is only required for malignant forms of CS, as their benign counterparts do not recur after surgical excision.

Contributors PP: Primary and corresponding author, responsible for background research into topic and literature review. Involved in collection and analysis of findings, involved in writing discussion of case study. RT: Surgical registrar involved in operation, involved in writing case presentation and discussion. Responsible for patient follow-up. SN: Pathologist involved in case, responsible for analysing pathology slides and confirming diagnosis. Created histology slides used in figure 4.

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