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CLINICAL CASE

A thoracic parasternal granular cell tumor in a child: About one case and review of the literature, discussion about treatment guidelines, is complete resection compulsory?



Tumeur à cellules granuleuses thoracique chez l'enfant : à propos d'un cas, discussion sur la prise en charge thérapeutique, l'exérèse complète est-elle indispensable ?

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Summary Granular cell tumor is a well known soft tissue tumor, very rare in children; we describe here the first case of GCT in this particular location in a child. The diagnostic is easily done with histopathology. The treatment is based on the complete resection, no other validated treatment exists. We reviewed the literature to find out if it would be safe to consider a simple follow-up after partial resection of the tumor.

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MOTS CLÉS

Tumeur à cellules
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Thoracique ;
Enfant ;
Traitement ;
Malignité

Résumé La tumeur à cellules granuleuses est une tumeur des tissus mous bien connue, atteignant très rarement les enfants. Nous décrivons le premier cas de tumeur à cellules granuleuses thoracique chez un enfant. Le diagnostic de cette tumeur est fait aisément par l'examen anatomopathologique. Le traitement est basé sur l'exérèse complète de la tumeur, il n'existe pas d'autre traitement validé. L'exérèse complète de la tumeur est parfois très délabrante, comme cela s'est présentée pour notre cas. Étant donné la nature le plus souvent bénigne de ces tumeurs, nous avons envisagé de procéder à une simple surveillance après résection partielle, nous avons effectué une revue de la littérature afin de valider cette stratégie.

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Case

We present the case of a nine year-old Caucasian white-skinned girl who developed a thoracic granular cell tumor (GCT) in the parasternal area. The lesion had been slowly growing for a few years. It presented as a nodule located on the right parasternal region at the 3rd intercostal space level; it was palpable, non-tender and firm, subcutaneous without any skin lesion, it seemed attached to the deep structures (costal cartilage and bone) but independent from the dermis. A complete physical examination was otherwise normal, in particular in the oropharyngeal region, no lymphadenopathy was found and the patient had no significant past medical history. Ultrasound showed an oval subcutaneous solid mass, measuring 14 × 12 × 6 mm. There were no fluid, no cystic features and no hypervascularisation. Histological confirmation was needed, giving the size of the nodule; we directly performed an excision of the entire lesion. As the nodule was adherent to the 3rd intercostal muscle, the extirpation was done cutting through the muscle in macroscopic negative margins. The histological analysis made the diagnosis of GCT. Pathologists observed a proliferation of large cells with abundant cytoplasm mostly granular, regular nuclei highly chromatic, small and rarely vesicular. There was no mitosis, no atypia and no necrosis (Figs. 1–2). The cytoplasmic granules were strongly periodic-acid-Schiff (PAS)-diastase resistant. Immunohistochemistry was positive for S-100 protein.

Unfortunately microscopic margins were positive (Figs. 1–2). As surgical complete excision of the lesion

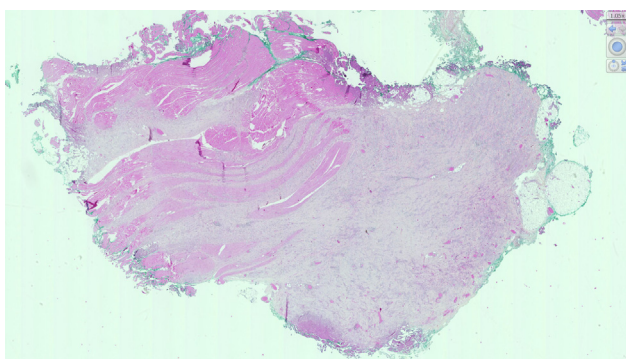


Figure 1 ($\times 1$) Photo of histology exam showing a poorly circumscribed tumor, infiltration of the adjacent muscle with incomplete excision.

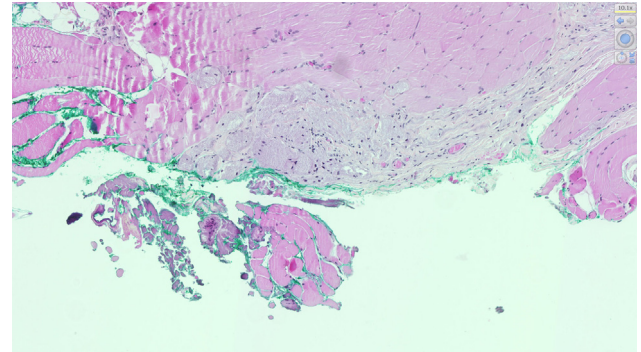


Figure 2 ($\times 10$) Photo of histology exam showing a poorly circumscribed tumor, infiltration of the adjacent muscle with incomplete excision.

would have been very damaging, we reviewed the literature for therapeutic options. No clear therapeutic guidelines exist. Other treatments such as chemotherapy or radiotherapy are not used in the case of a non-metastatic tumor. The tumor presented no clinical nor histological malignant characteristics, implying low risks of local recurrence and metastasis. Dermatologists, plastic surgeons and pathologists agreed for an attentive follow-up. After eighteen months of follow-up, there have been no clinical signs of recurrence (Fig. 3).

Discussion

Granular cell tumor (GCT) also called Abrikossov tumor is a rare mostly benign tumor originated from Schwann cells of peripheral nerves, first described in 1926 by the Russian pathologist Abrikossov. It can affect any organs or tissues but generally arises in the subcutaneous or cutaneous tissue of the head and neck region, the tongue and the larynx [1–3]. This tumor mainly occurs in 30 to 60-year-old women, more frequently in African Americans [2,4]. About 30 cases have been reported in children essentially in head and neck area, half with other anomalies (nervous, cardiovascular, endocrine and musculoskeletal), no associated congenital abnormality or genetic syndrome have ever been described [2].

It often presents as a non-tender, firm nodule, with a diameter inferior to 3 cm, asymptomatic or with mild itch or paroxysmal tenderness [2], other symptoms are related to the location.

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