Scholars Journal of Applied Medical Sciences
Abbreviated Key Title: Sch J App Med Sci
ISSN 2347-954X (Print) | ISSN 2320-6691 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Synovial Sarcoma of Parotid Gland: Very Rare Cause of Malignant Tumors of the Neck in Children

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DOI: 10.36347/sjams.2021.v09i05.010 | **Received:** 28.03.2021 | **Accepted:** 06.05.2021 | **Published:** 14.05.2021

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Abstract **Case Report**

Synovial sarcoma is an unusual malignant tumour, derived from soft tissue mesenchymal cells,It arises most commonly in the deep soft tissues of lower extremities, especially in the region of the lower thigh, with a predilection for sites in proximity to large joints, such as the knee and ankle. Only 3% to 10% of all synovial sarcomas occur in the head and neck region. Primary synovial sarcoma of the parotid gland is exceptionally uncommon, with few cases reported in the literature. We report the case of a 10-month-old infant, who consulted for right parotid swelling evolving for 1 month, rapidly increasing in size without inflammatory signs or deterioration of the general condition. Ultrasound, CT and MRI were done showing a locally infiltrating lesion process. The infant underwent a biopsy with immunohistochemical study confirming the diagnosis of synovial sarcoma of the right parotid gland.

Keywords: synovial sarcoma; parotid gland, imaging.

Abbreviations: MRI: magnetic resonance imaging, CT: computed tomodensitometry.

INTRODUCTION

Neck masses are common findings in pediatric patients, and are benign in the majority of cases, in addition to the more common tumors (lymphoma, rhabdomyosarcoma), rare tumors are also encountered in the pediatric head and neck tumors(1).

Synovial sarcoma is an unusual malignant tumour, derived from soft tissue mesenchymal cells, It arises most commonly in the deep soft tissues of lower extremities, especially in the region of the lower thigh, with a predilection for sites in proximity to large joints, such as the knee and ankle [2]. It typically occurs in young adults with sex ratio of 2/1 [3, 4]. Only 3% to 10% of all synovial sarcomas occur in the head and neck region, the pharyngeal and cervical areas are most commonly affected while the larynx is the least common site [5].

Primary synovial sarcoma of the parotid gland is exceptionally uncommon, with few cases reported in the literature. A diagnosis of Synovial sarcoma was established based on the histology [6]. Surgery is the treatment of choice, given that the role of adjuvant therapy is controversial [7].

CASE REPORT

A 10-month-old infant who consulted for right parotid swelling evolving for 1 month , rapidly increasing in size without inflammatory signs or deterioration of the general condition (figure 1) . The biological assessment was normal. The ultrasound scan showed a solid-cystic mass at the expense of the right parotid gland, not clearly limited, vascularized in color Doppler study (Figure 2). The CT showed a solid-cystic mass centered on the right parotid space with signs of hemorrhage, enhanced after injection of the contrast product which fills the retrostylar space and partially fills the prestylian space and the right parapharyngeal space, it also showed the encompassing of the right internal carotid artery over a circumference> 180 ° which has remained patent (figure 3). On MRI this mass had a heterogeneous signal on T1 and T2 weighted, restricted diffusion, heterogeneously enhanced after injection of gadolinium delimiting large areas of necrosis (figure 4). The infant underwent a biopsy with immunohistochemical study confirming a diagnosis of synovial sarcoma of the right parotid gland: the histological study showed a tumor proliferation of both fusocellular and epithelial contingents. The immunohistochemistry using immunoperoxidase staining, after heat-induced epitope retrieval, revealed a moderate and diffuse membrane

expression of epithelial tumoral cells of the anti-EMA antibody and an intense nuclear expression of 35% of tumoral cells of the anti-Ki67 body. Further, reverse-transcription PCR for t(X; 18) (p11; q11) has been proven.

The infant underwent abdominal and pelvic CT as an extension assessment that did not show any secondary location. After a multidisciplinary consultation meeting, the management consisted of a neoadjuvant chemotherapy for tumor volume reduction, then a subsequent surgical resection.

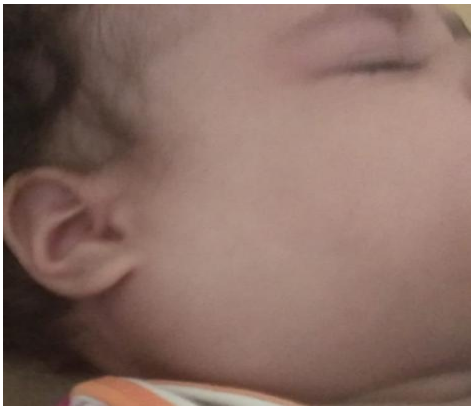

Fig- 1: Right parotid swelling without inflammatory signs

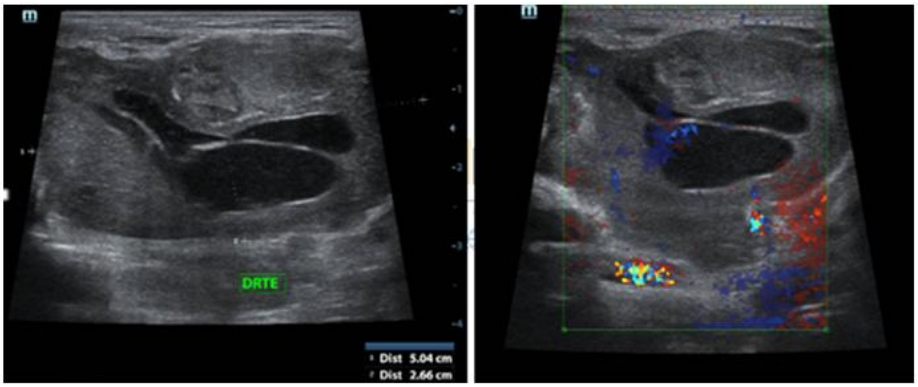

Fig-2: solid-cystic mass at the expense of the right parotid gland, not clearly limited, vascularized in color Doppler study.

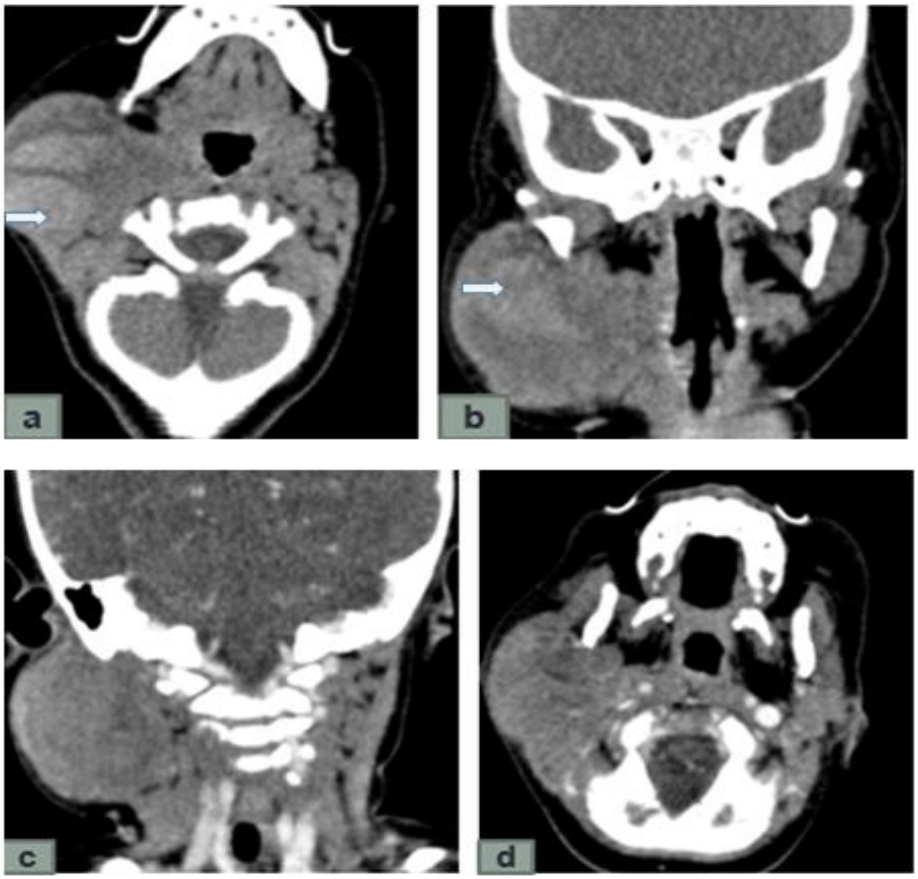

Fig- 3: CT without (a-b) and with contrast (c-d): solid-cystic mass centered on the right parotid space with signs of hemorrhage (arrows), enhanced after injection of the contrast product.

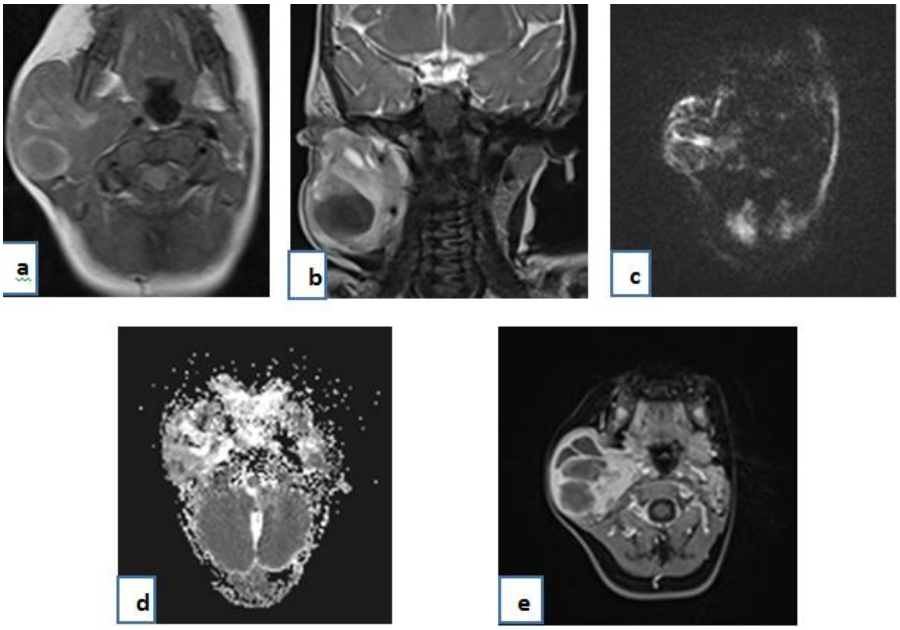

Fig- 4: MRI showing a heterogeneous signal on T1 (a) and T2 weighted (b), restricted diffusion (c-d), heterogeneously enhanced after injection of gadolinium delimiting large areas of necrosis(e).

DISCUSSION

Synovial sarcoma (SS) is a malignant mesenchymal tumour, predominantly found in the deep soft tissues of lower extremities, whereas only 3% occur in the head and neck region [2], The most common site in head and neck is the hypopharynx, with the larynx being the least common [6]. Primary synovial sarcoma of the parotid gland is exceptionally uncommon. The origin of these tumours is believed to be from an undifferentiated mesenchymal cell that undergoes synovioblastic differentiation [7]. Despite its name, synovial sarcoma is not restricted to periarticular sites [8]. According to Leader et al [9], it is currently accepted that synovial sarcoma arises from undifferentiated or pluripotent mesenchymal cells with dual differentiation capacity, both epithelial and mesenchymal. This is why this Author suggests classifying synovial sarcoma as carcinosarcoma. This theory is in contrast with others describing a clear link with joints as beginning sites of these sarcomas. The peak incidence is in adolescents and young adults between 16 and 49 years of age (median 34 years), and the tumor affects predominantly males [10] .To the best of our knowledge, this is the youngest recorded patient with synovial sarcoma of the parotid gland.

A painless neck mass was the single most common presenting symptom [11] and the long duration of symptoms may simulate a benign process [12]. On ultrasound this tumor appears as a solid, round or lobulated, hypoechoic and heterogeneous soft-tissue mass, Hypoechoic areas representing hemorrhage or necrosis may occur [12]. On CT the tumor is isodense or slightly hypodense to muscle, with areas of low attenuation representing necrosis or hemorrhage. Synovial sarcoma frequently demonstrates a multinodular appearance, and heterogeneous enhancement, Calcifications may occur [12]. MRI shows a heterogeneous, well-defined, multilobulated soft-tissue mass with signal intensity similar to or slightly higher than muscle on T1-wheighted imaging (WI), and high signal intensity on T2-WI. The signal heterogeneity has been termed the "triple sign", represented by intermixed areas of high, intermediate and low signal intensity, and representing areas of hemorrhage or necrosis, solid tumor, and calcified or fibrotic regions [1]. Intervening septa are frequently present, as well as fluid levels. Prominent, heterogeneous and early enhancement is usually present [12]. A diagnosis of Synovial sarcoma was established based on the histology and, in particular, the positivity for AE1/AE3, CK8/18, EMA, and CD99 and the strong reaction to CD56 and TLE1. Recently, reverse-transcription PCR or FISH for t(X; 18) (p11; q11) was necessary to prove a diagnosis of Synovial sarcoma [6].

The optimal treatment of these tumours seems to be multimodal. Radical surgery represents the first approach. Post-operative radiation treatment has been found to improve the prognosis in head and neck localizations rather than in the extremities [9, 13]. Surgery followed by radiotherapy with or without chemotherapy has been advocated by most studies. However, there is no evidence that support the sole treatment modality of chemotherapy or radiotherapy [6]. Recently, some Authors suggested the possible role of epidermal growth factor receptor (EGFR) and human epithelial growth factor receptor 2 (Her-2/neu) in the cancerogenesis of synovial sarcoma thus suggesting that the anti-EGFR monoclonal antibody may play a role in the therapeutic approach [14, 15] (.No standardized treatment protocol for now is available for this type of tumor.

Despite aggressive surgical resection, local recurrence occurs in up to 50% of patients and metastatic disease develops in 41%, to the lung, lymph nodes and bone [12]. Follow-up should be lifelong due to the tendency for late recurrences and metastasis [6].

CONCLUSION

The primary synovial sarcoma of the parotid gland is extremely rare, especially in children, the diagnosis and clinical management can be a challenge.the diagnosis is based on the imaging, confirmed by the histological, immunohistochemical and molecular study, surgery is necessary while the place of radiochemotherapy remains controversial.

Conflict of Interest:

All authors state that they have no conflicts of interest.

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