

Primary cutaneous alveolar rhabdomyosarcoma: Case report in a young adult

Rabdomiossarcoma cutâneo primário de padrão alveolar: relato de caso em adulto jovem

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RESUMO

Sarcomas são neoplasias mesenquimais malignas, raras, que acometem, principalmente, crianças e adolescentes. O rabdomiossarcoma, subtipo oriundo da musculatura esquelética, é condição incomum em adultos, acometendo sítios de localização não habitual, crescimento rápido e de difícil tratamento. Apresenta-se caso de adulto jovem com nodulação em lóbulo auricular esquerdo, cuja análise histopatológica e imuno-histoquímica confirmou tratar-se de rabdomiossarcoma alveolar, o qual foi conduzido em conjunto com a Oncologia.

Palavras-chave: Rabdomiossarcoma Alveolar; Miogenina; Desmina; Excisão de Linfonodo

ABSTRACT

Sarcomas are rare and malignant mesenchymal neoplasms that mainly affect children and adolescents. Rhabdomyosarcoma, a subtype originating from skeletal muscle, is an uncommon condition in adults. It affects sites of unusual location, presents fast growth, and is challenging to treat. We report a case of a young adult with nodules in the left auricular lobe. The histopathological and immunohistochemical analysis confirmed the alveolar rhabdomyosarcoma, and treatment was conducted in association with Oncology.

Keywords: Rhabdomyosarcoma, Alveolar; Myogenin; Desmin; Lymph Node Excision

INTRODUCTION

Rhabdomyosarcoma is an uncommon malignant neoplasm occurring mainly in children. It affects most commonly the head and neck (35%), genitourinary system, and extremities (40%). Less frequently, the condition affects the trunk, orbit, intrathoracic region, and retroperitoneum. The alveolar subtype is the most aggressive, with the worst prognosis.

CASE REPORT

A twenty-one-year-old man reported a lesion on the left ear lobe for about a month, with a progressive increase in size. He denied other associated symptoms such as pain, fever, or weight loss. The general physical examination was normal.

Case Reports

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FIGURE 1:
Nodulation
of hardened
consistency in
the ear



FIGURE 2: Detail of
painless nodulation

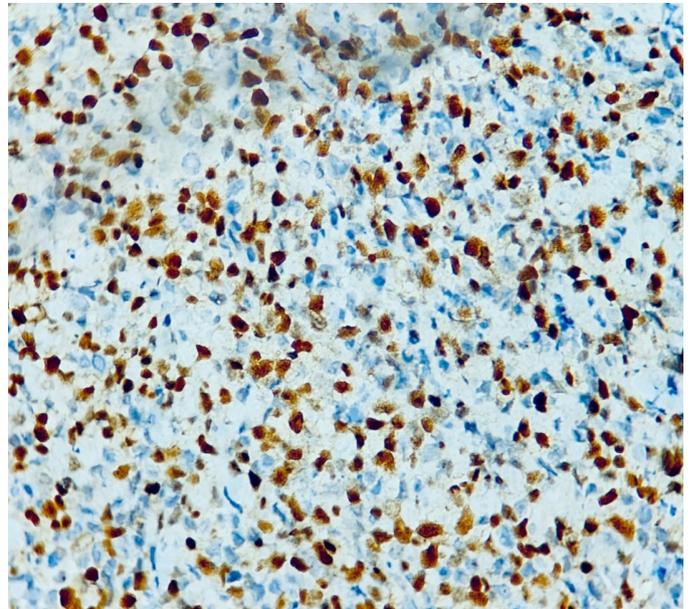


FIGURE 3: Resection of the left ear tumor, resection of a left parotid lesion,
and left modified radical cervical lymphadenectomy

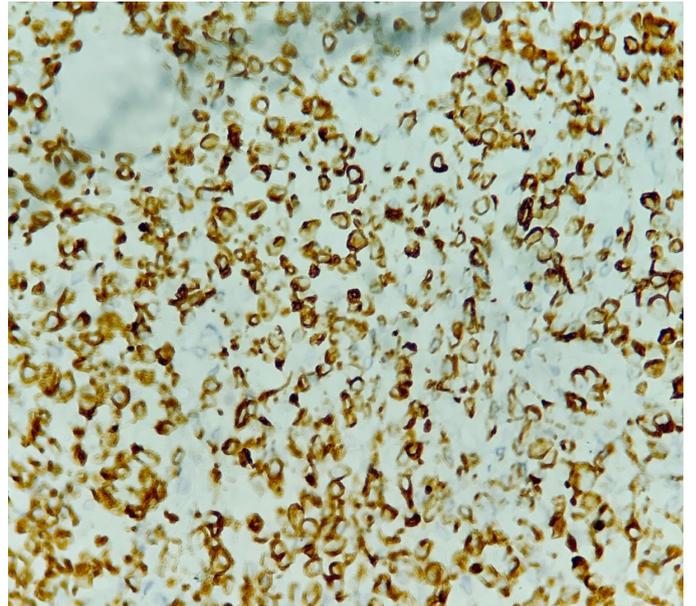


FIGURE 4: Neoplasm of small and round cells infiltrating adipose tissue.
Positive immunohistochemistry for desmin and myogenin

The dermatological examination revealed presence of hardened nodulation, painless on palpation, and without phlogistic signs at the anterior region of the left lobe, extending to the posterior region (Figures 1 and 2). An attempt to perform fine-needle aspiration (FNA) biopsy was unsuccessful. After an incisional biopsy, the anatomopathological examination showed neoplasia of small and round cells, infiltrating adipose tissue.

The immunohistochemistry was positive for desmin and myogenin (Figures 3 and 4), confirming the diagnosis of alveolar rhabdomyosarcoma. The patient was then referred to the On-

ology Service. Computed tomography of the neck to detect metastasis evidenced an intraparotid lymph node enlargement on the left, with suspicion for secondary neoplastic involvement. Therefore, we conducted the resection of the left ear tumor, left parotid lesion, and left modified radical cervical lymphadenectomy (Figure 5). Histopathological analysis of tumor resection showed alveolar rhabdomyosarcoma, stage pT3 pN1. The resection product from parotid lesion showed infiltration by such sarcoma. On the left cervical lymphadenectomy, rhabdomyosarcoma metastasis was confirmed in four of the thirty resected lymph



FIGURE 5: - Positive immunohistochemistry for desmin and myogenin: alveolar rhabdomyosarcoma

nodes. The patient underwent four chemotherapy sessions and 28 radiotherapy courses and is currently being followed up.

DISCUSSION

Sarcomas are rare malignant mesenchymal neoplasms that mainly affect children and adolescents.

Rhabdomyosarcoma, a subtype originating from skeletal muscle, is the most common type of soft tissue sarcoma in children, corresponding to approximately 50% of these tumors. The head and neck region are the most affected site, followed by the genitourinary tract, extremities, chest, and retroperitoneum.¹ There are few reports in the literature of primary cutaneous rhabdomyosarcoma since this is an extremely rare condition. In adults, the occurrence is even more unusual – about 90% of all rhabdomyosarcomas occur in individuals under 25 years old – affecting sites of unusual location, presenting fast growth, and being difficult to treat.²

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A biopsy is necessary to conclude the diagnosis. The biopsy can be surgical or with thick or fine needle aspiration (FNA).

Differential diagnoses of cutaneous lesions of the head and the neck include: hemangioma, lymphoma, lymphangioma, leukemia cutis, angiofibroma, neuroblastoma, cutaneous myofibroma, glioma, cellulitis, abscess, mastoiditis, and other sarcomas.²

A dermal infiltrate of small and oval-shaped cells with an acidophilic cytoplasm characterizes the histopathology of cutaneous alveolar rhabdomyosarcoma. Such changes can occur in various pathological conditions, and immunohistochemistry is essential to define the diagnosis. It is possible to find rhabdomyoblasts and characteristic neoplastic multinucleated giant cells in well-differentiated types, although immunohistochemistry is always an integral part of the diagnosis.²

There are four histopathological types, with their variants: embryonal, alveolar, pleomorphic, and sclerosing. The alveolar subtype is the most aggressive, with rapid progression, which causes early metastasis and increasing mortality rates. In this histological type, there is no association with environmental factors. Chromosomal translocations – t (2;13) and t (1;13) – are fundamental pieces in its development. There are also reports of involvement of individuals with other changes in the central nervous system, urogenital and gastrointestinal tract, and melanocytic nevi; thus it is believed these groups have to have a genetic predisposition.¹

The first choice treatment is surgical, always combined with adjuvant chemotherapy to avoid metastasis. Radiotherapy is necessary when total lesion resection is not possible.

The prognosis depends on the site of origin, size of the lesion, clinical stage, age of the patient, and histological type. Good prognostic factors include early age at diagnosis, primary site in the genitourinary tract and orbit, and embryonal and botryoid histological types.^{2,3}

The totally resected lesions are associated with a 90% survival in five years, which evidences the importance of diagnosis and treatment institution as early as possible.^{2,4} ●

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