CASE REPORT

Alveolar rhabdomyosarcoma of nasal presentation: Case report

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Abstract Sarcoma is a malignant tumor originating in mesenchymal primitive cells that in normal circumstances develops in supportive tissues such as muscle and bone. There are three rhabdomyosarcoma variants according to its histological presentation: Embryonal, alveolar, and pleomorphic with alveolar rhabdomyosarcoma being the most aggressive. The case is presented of an 8-year-old male patient who started with a progressively growing left nasal wing 3-4 mm mass with no color change diagnosed as an alveolar rhabdomyosarcoma with pathology documenting a small round blue cell tumor with positive immunohistochemistry to desmin and myogenin. Skull and face magnetic resonance, chest computed tomography, and bone marrow aspirate were negative for disease extension. The lesion was macroscopically resected at 100%, with pathology reporting isolated tumor foci, with microscopic residual tumor on lateral margins, and it is therefore finally classified as E II. This is a very rare neoplasm with an unusual presentation, with only 4 cases reported in the literature, hence, the importance of taking into account this diagnostic possibility as well as the knowledge on how to approach it.

KEY WORDS
Alveolar rhabdomyosarcoma; Nasal; Multidisciplinary; Sarcoma; Pediatric age

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INTRODUCTION

Sarcoma is a malignant tumor originating in primitive mesenchymal cells that under normal circumstances develops in supportive tissues such as muscle and bone. Since skeletal muscle cells are present in most part of the body, this malignancy can occur on almost any anatomic location1.

There are three variables of rhabdomyosarcoma according to its histological presentation such as the embryonal type with its variants botryoid and spindle-shaped cells, alveolar (including the solid alveolar variant), and pleomorphic. Alveolar rhabdomyosarcoma is the most aggressive, it occurs more commonly in adolescents and young adults, and accounts for 20% of all rhabdomyosarcoma cases, generally with onset on the torso and limbs2.

Annual incidence of rhabdomyosarcoma in 20-year-old or younger individuals is 4.3 cases per millions of children, with approximately 350 new cases diagnosed in the US every year. Of the tumors occurring in head and neck structures, almost 40% correspond to rhabdomyosarcomas3.

Anatomically, they are classified as parameningeal, orbital, nonparameningeal, and nonorbital; parameningeal sites include the nose, paranasal sinuses, nasopharynx, middle ear, mastoid process, infratemporal fossa, and pterygopalatine fossa4. In nearly 90% of cases, chromosomal translocations are appreciated in genes PAX3 (2q33) FKHR/ALV (13q14), and less commonly in PAX7 (1p36) KHR/ALV (13q14)5.

The purpose of the present article is to describe the case of a patient with Stage II nasal alveolar rhabdomyosarcoma, which is a rare neoplasm in our country and worldwide and worthy of a comprehensive literature review with regard to multidisciplinary treatment, to begin with, the participation of radiologists, well-trained pathologists familiarized with pediatric tumors and surgeons as well as oncologists specialized in the treatment of this neoplasm6.
2.5 cm × 2 cm with even color to the boy’s skin and with 60% reduction in volume (Fig. 5). On July 21, 2015, the lesion was 100% macroscopically resected (Fig. 6), but the pathology report indicated isolated tumor foci with lateral margins microscopic residual tumor, and the lesion was finally classified at Stage II. Owing to the permanence of residual disease, and to the fact that it was an aggressive histological variety, the patient was referred for radiotherapy to the primary site.

He received CT with weekly VIE alternating with VAC every 3 days for 3 weeks and VAC + IF and EF every 3 weeks twice with the treatment concluding on July 21, 2016.

DISCUSSION

Rhabdomyosarcoma is an extremely rare malignant tumor that accounts for 0.4-1.0% of all soft tissue sarcomas. These tumors usually occur in adolescents or young adults’ lower limbs with rhabdomyosarcoma of the nasal or paranasal region even being more unusual7,8. Rhabdomyosarcomas are generally divided in meningeal and parameningeal sites with parameningeal sites including the middle ear, nose cavity, paranasal sinuses, nasopharynx, and the infratemporal fossa10. There are only 4 cases reported in the literature, with this malignancy being more common in females than in males at a 2:1 ratio8.

In general, it is a slowly growing, well-vascularized mass, with no clinical characteristics suggesting malignant disease. When it occurs in the nasal cavity, the nasal obstruction can be the only symptom11.

Different histological types of rhabdomyosarcoma of head and neck region have been identified, including embryonal, alveolar, and pleomorphic rhabdomyosarcoma2-9. Embryonal and alveolar patterns are the most common. Histologically, embryonal rhabdomyosarcoma is comprised by primitive round spindle-shaped cells with rhabdomyoblasts, whereas alveolar rhabdomyosarcoma consists of malignant cells grouped in fibrovascular septae that form alveoli-like spacings10.

Poorly differentiated rhabdomyosarcomas can be difficult to distinguish from poorly differentiated Ewing sarcoma or neuroblastoma. Tumors with a high predominance of fusiform cells can be confused with leiomyosarcomas, fibrosarcomas or malignant fibrous histiocytomas. Immunohistochemically, intermediate filaments specific to muscle cells such as myoglobin, myosin, desmin, and creatine kina-
se MM isoenzyme are regarded as rhabdomyosarcoma-specific markers12.

Microscopically, tumor cells tend to be smaller and rounded, offering a densely cellular appearance, and they are often included in the round, blue cells classification, where malignant lymphoma, neuroblastoma, and synovial sarcoma (biphasic) often stand out as differential diagnoses10.

In spite of tumor slow growth, the prognosis is generally poor11 and depends on a combination of patient age, histological nature, clinical stage, and tumor location 11, with a high tendency toward early metastatic spread11. Prognostic variables identification depends on different groups of patients with excellent, very good, fair, and poor prognosis; on the site (favorable vs. unfavorable, with the orbit being the most favorable site); surgical respectability (Groups I and II vs. Group III); histology (embryonary vs. alveolar); and age, with intermediate grade alveolar rhabdomyosarcoma showing 40-50% prognosis1.

CONCLUSIONS

The article represents a case report of an alveolar rhabdomyosarcoma nasal presentation. We might conclude that the approach should be multidisciplinary and individualized.

DECLARATION INTEREST

The authors declare not having any conflicts of interest.

REFERENCES