Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Medicine

Unusual Extraosseus Location of Ewing's Sarcoma: Case Report

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DOI: 10.36347/sjmcr.2024.v12i05.094 | **Received:** 21.04.2024 | **Accepted:** 25.05.2024 | **Published:** 29.05.2024

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Abstract **Case Report**

Introduction: Extraskeletal Ewing sarcoma is a very rare malignant tumor. The head and neck region is an unusual primary location for this type of tumor [1]. Clinical case: We present a 13-year-old girl with no known personal history. She was brought for consultation in June 2023 presenting a tumor mass in the left nasal fossa that caused slight nasal obstruction. She reported episodes of epistaxis and left epiphora, and denied any history of trauma to the facial region or some type of viral or catarrhal infection. During the physical examination we found a solid, elliptical mass between the body of the malar bone and the anterosuperior part of the left maxillary bone. There was a boundary between the lesion and the surrounding muscle and it did not infiltrate the skin. ***Discussion***: Ewing sarcoma is the second most common malignant bone neoplasm in pediatrics; however, it is uncommon in soft tissues with a low incidence rate of 1.1%, including sinonasal SE, not to mention primary neuroectodermal tumors, which are tumors of small, round cells derived from soft tissue, and which belong to the Ewing sarcoma family [6]. ***Conclusion***: Ewing's sarcoma is a rare tumor; it usually occurs in children and young people with highly aggressive behavior and spreads rapidly and extensively. The presence of small round blue cells plus the immunohistochemical study confirms the diagnosis. For treatment there is still no consensus and universally accepted protocols, however, a multidisciplinary approach is required, with an aggressive approach combined with surgery and radio-chemotherapy. The good general condition of the patient at the time of diagnosis was very favorable in his response to oncological treatment and evolution; one year after diagnosis and initial management, no clinical or imaging signs of tumor persistence or recurrence were found.

Keywords: Ewing Sarcoma, Extraosseous, Chemotherapy.

INTRODUCTION

Extraosseous Ewing's sarcoma is one of the rare tumors of the head and neck. It was described for the first time in 1918 by James Ewing, but it was not until 1921 that its histogenetic origin was described [1]. They are very aggressive tumors that cause early metastasis. Therefore, it forces a very unfavorable prognosis to be given; they are part of a large family of tumors whose origin is found in embryonic cells; ES shares characteristics with primitive neuroectodermal tumors [2, 3]. It normally develops in muscle and bone tissues, but can develop in any location [4]. It is found within small, round and blue cell tumors, along with other tumors, such as lymphomas, primitive neuroectodermal tumors, rhabdomyosarcoma, neuroblastoma, among others, which we must think about as a differential diagnosis [5]. This type of tumor appears most frequently in children and young adults, many authors indicate between 8 and 20 years of age, but they can occur at any age with a predominance in males in a ratio of 1.5:11 [6]. Prognosis depends on the existence of metastasis because Ewing sarcoma is highly malignant and causes metastasis to the bone and lung, so survival is relatively low [7].

With this article we intend to describe the case of a girl with sinonasal Ewing Sarcoma, the head and neck region is an unusual primary location for this type of tumor.

CLINICAL CASE

We present a 13-year-old girl with no known personal history. She was brought for consultation in June 2023, presenting a tumor mass in the left nostril that

caused slight nasal obstruction. She reported episodes of epistaxis and left epiphora, and denied any history of trauma to the nasal cavity. Facial region or some type of viral or catarrhal infection. During the physical examination we found a solid, elliptical mass between the body of the malar bone and the anterosuperior part of the left maxillary bone. There was a boundary between the lesion and the surrounding muscle and it did not infiltrate the skin (Fig. 1).

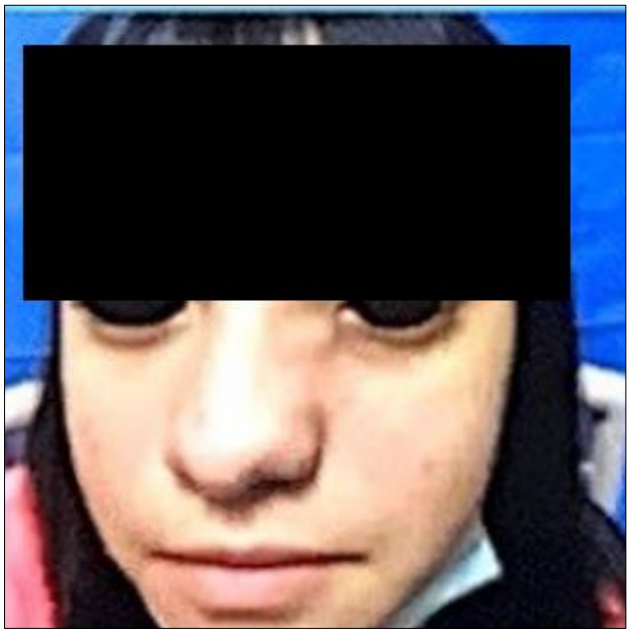

Figure 1: tumor at the level of the nasion that extends to the inner canthus.

Once the physical examination and complementary laboratory tests were performed, we indicated a craniofacial mass tomography (TC) and a magnetic resonance imaging (RMN). The report showed us the presence of a tumor mass in the left nasal fossa, heterogeneous with bone erosion to structures. Bony neighbors.

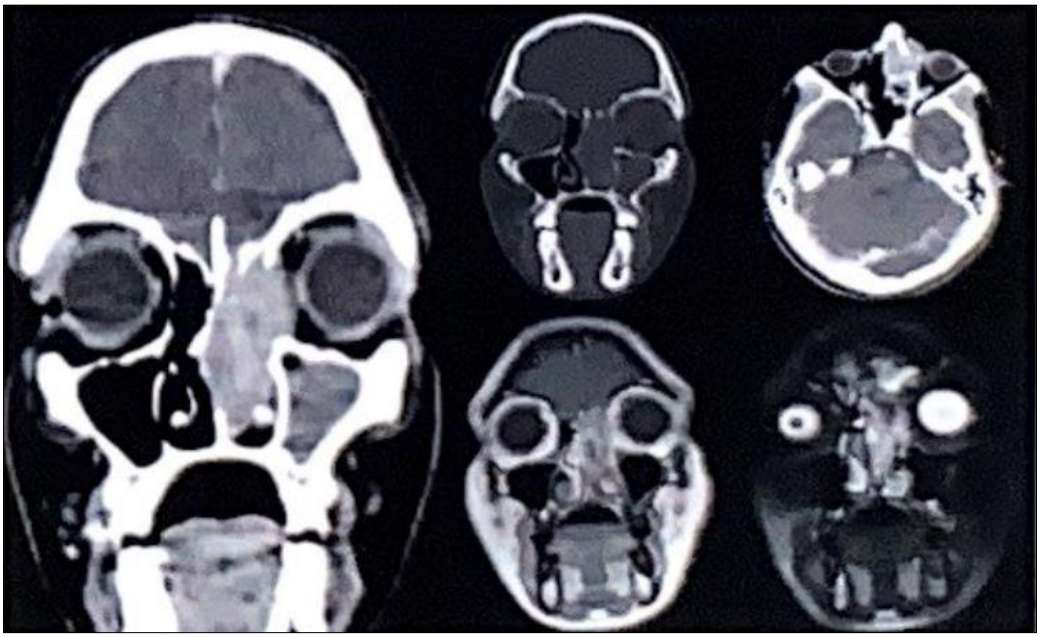

Figure 2: Tomography of the craniofacial mass (MRI), coronal and axial section. Magnetic resonance imaging (RMN)

Figure 3: Bone scan

RESULTS

We performed incisional surgery to take a biopsy of the left nasal fossa (Fig. 3). We found a rounded tumor mass, with solid elements of a hard consistency, and friable towards the implantation area, consisting, for the most part, of a compact proliferation of small cells.

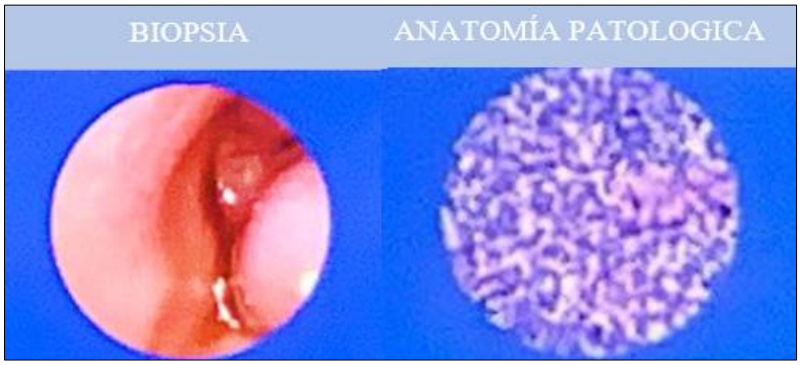

Figure 4: Incisional biopsy of the right nasal fossa: rounded tumor, with solid elements of hard consistency, and friable towards the implantation area

The incisional biopsy reported rhinosinusal Ewing sarcoma. In addition, tumor markers were performed to ensure the diagnosis. Dako CD 45 clone 2b11 plus Pd7/26 was performed to rule out tumors of a lymphoid nature, which was negative. Dako CD99 and anti- synaptophisine antibodies were performed (Dako). Clone 19) looking for the presence of epithelial tumors and neuroblastomas which was positive, it was interpreted as Ewing's sarcoma of the nasal cavity. referred to oncology where she began treatment protocol 14 cycles of chemotherapy with etoposide, vincristine, actinomycin D, ifosfamide and adramycin + IMRT.

Three months after the treatment we found favorable positive changes with good evolution. During the physical examination we found no signs of aggression or bone lysis. We indicated monthly evolutionary monitoring.

DISCUSSION

Ewing's sarcoma is the second most common malignant bone neoplasm in pediatrics; however, it is uncommon in soft tissues with a low incidence rate of 1.1%, including sinonasal SE, not to mention primary neuroectodermal tumors, which are tumors of small, round cells derived from soft tissue, and which belong to the Ewing sarcoma family [6].

The diagnosis is based on clinical, radiological and pathological elements. Symptoms and signs are variable, frequent epistaxis may appear, especially in advanced stages of the disease, nasal obstruction,

purulent rhinorrhea, if there is lysis or bone destruction, the patient may report facial pain, our patient presented unilateral left nasal obstruction and epistaxis. Histologically, Ewing sarcoma has both mesodermal and ectodermal origin, therefore its classification is difficult. It is among the small round cell tumors of childhood, characterized by the presence of small round cells with hyperchromatic nuclei, well-defined borders and absence of intercellular material; There is abundant cytoplasmic glycogen [7]. The exact etiology remains unknown, some authors propose that this tumor presents a cytogenetic anomaly that differentiates it from other pediatric tumors such as neuroblastoma and rhabdomyosarcoma, not related to congenital syndromes or neoplastic syndromes; The reviewed literature mentions that this family of sarcomas shares a single specific translocation sequence involving chromosomes 11 and 22, t (11;22) (q24;q12), which results in the expression of a chimeric protein, EWSR1. -FLI1. This translocation is present in 85-95% of cases that have presented tumors of the Ewing Sarcoma family [8]. Tumor markers are essential to be able to make differential diagnosis with other tumors that share characteristics similar to ES; It is necessary to evaluate the immunohistochemistry of Vimentin when it is positive it speaks in favor of mucosal melanoma and rhabdomyosarcoma, to rule out neuroendocrine carcinoma , rhabdomyosarcoma and pituitary adenoma, CD56 markers are indicated, positive in these cases; The S100 protein is positive up to 30% of the SE, in addition to CD99, which is considered the most sensitive marker , due to a strong and diffuse CD99 membranous reactivity of Ewing Sarcoma [9]. The differential diagnosis of tumors in the nasal cavity and paranasal sinuses with or without intracranial involvement includes meningiomas or metastases, direct extension of tumors from the base of the skull or the nasopharynx, ruling out esthesioneuroblastoma , nasopharyngeal carcinoma, melanoma, rhabdomyosarcoma or lymphoma, sometimes including nasal angiofibroma [10].

The most common characteristics at the level of computed tomography is the presence of a mass with soft tissue density that captures contrast medium in an irregular manner; and at MRI level, lesion isointense or slightly hyperintense on T1 and hyperintense on T26. Findings that we found in the MRI performed on our patient [9]. In the literature consulted, treatment for the Ewing family of tumors is based on the administration of a high dose of chemotherapy for systemic control of the disease, followed by local control, which includes extensive resection or irradiation of the tumor. Affected tissue. Due to the aggressiveness of these tumors, a combined treatment is proposed, consisting of surgical resection with adjuvant chemo and radiotherapy. Neoadjuvant chemotherapy can also be used to facilitate surgical resection and reduce intraoperative risks. Chemotherapy is very relevant, since it produces a significant improvement in the prognosis of these patients, guaranteeing survival [8-10]. The EuropeanInter group Cooperative recommends 14 cycles of etoposide, vincristine, ifosfamide, and adriamycin. According to this protocol, chemotherapy is repeated every 3 weeks. Since the introduction of chemotherapy, survival has improved significantly, with a survival of 5 years. The most frequent sites of metastasis correspond to the lung, liver and bone marrow [10].

CONCLUSION

Ewing's sarcoma is a rare tumor; it usually occurs in children and young people with highly aggressive behavior and spreads rapidly and extensively. The presence of small round blue cells plus the immunohistochemical study confirms the diagnosis. For treatment, there is still no consensus and universally accepted protocols, however, a multidisciplinary approach is required, with an aggressive approach combined with surgery and radio-chemotherapy. The good general condition of the patient at the time of diagnosis was very favorable in his response to oncological treatment and evolution; one year after diagnosis and initial management, no clinical or imaging signs of tumor persistence or recurrence were found.

Conflicts of Interest: The authors declare that there is no conflict of interest regarding the publication of Este paper.

Financing: Self-funded

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