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Pleomorphic Rhabdomyosarcoma in Adult: A Very Uncommon Occipital Location

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Abstract

Rhabdomyosarcome is a commun tumor in children with all its types (pleomorphic, embryonal, alveolar and spindle cell rhabdomyosarcoma). However, it is a very rare tumor in adults, the commun type is the pleomorphic rhabdomyosarcoma but it is very rare in the head and neck region.

The diagnosis of the pleomorphic RMS is made by the histopathologic study, this shows the interest of a biopsy before the surgical procedure. The treatment of patients with malignant mesenchymal tumors of the head and neck region is multidisciplinary (radical surgery with postoperative radiotherapy and or chemotherapy) with poor prognosis.

Keywords: Pleomorphic Rhabdomyosarcoma; Adult; Occipital Region

Introduction

Rhabdomyosarcoma is one of the most common tumors of childhood and adolescence, (more than 50% of soft tissue sarcomas) [1]. However, it is a very rare tumor in adults (less than 3% of adult soft tissue sarcomas) [2]. There are four major categories of rhabdomyosarcoma, which include pleomorphic, embryonal, alveolar and spindle cell rhabdomyosarcoma [3]. The pleomorphic forme is the most frequente in adults but the neck localisation is the rarest. To our knowledge, there are no previous case reports of adult pleomorphic rhabdomyosarcoma occurring in the occipital region in the literature. In this article we will report an uncommun case of pleomorphic rhabdomyosarcoma occurring in the occipital region.

Case Report

A 72-year-old male patient with no medical history of surgery or trauma, was referred to our department for a huge occipital mass (Figure 1) evolving over 10 months and gradually increasing in volume. The clinical examination found an occipital swelling overflowing on the neck without any inflammatory signs in the skin, the tumor was firm on palpation , painless, well limited, about 8 cm major axis. Examination of cervical lymph nodes showed no palpable lymphadenopathy. The rest of the maxillofacial, neurological and somatic examination were without particularities. A cranio-facial CT scan (Figure 2) showed the presence of a left-sided, heterogeneous occipital tissue overflowing on the nucal region, containing areas of central necrosis, measuring 59mm x 73mm x 92mm, that enhanced after contrast injection. The mass was interposed between the spinous muscle and the levator scapulae muscle. The tumor does not infiltrate the skin from which it is separated by subcutaneous fat. The CT scan showed no lymphadenopathy associated.

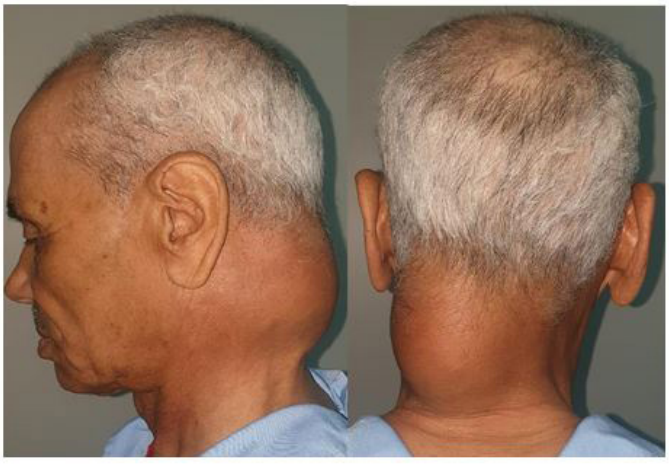

Figure 1: occipital mass overflowing on the nucal region

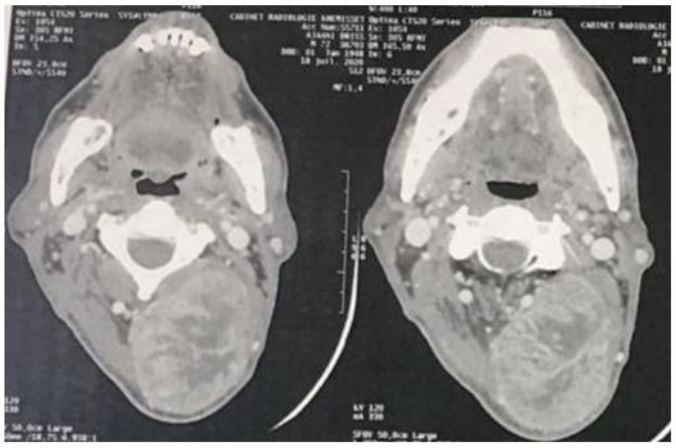

Figure 2: CT scan showing a heterogeneous occipital tissue containing areas of central necrosis

Under general anesthesia, we started with intra-operative frozen section that showed pleomorphic cells with enlarged hyperchromatic nuclei and eosinophilic cytoplasm. The surgery was completed by a large excision of the tumor limited by the cervical spine.

A histopathological examination of the surgical specimen showed pleomorphic cells with enlarged hyperchromatic nuclei and eosinophilic cytoplasm.

Immunohistochemical testing detected neoplastic cells expressing desmin, myogenin, CK AE1/AE 3, EMA, AML, H-caldesmone, CD 34, CD 31, KI67, and not expressing the HMB45, Melan A and the PS100; confirming the diagnosis of pléomorphic rhabdomyosarcoma.

Computerized tomography (CT) examinations of the abdomen and thorax revealed no metastases, and no lymphadenopathy was detected in the neck.

The follow-ups were simple, the patient underwent adjunctive chemotherapy to eradicate any residual tumours.

Discussion

Rhabdomyosarcome is a commun tumor in children with all its types. About 30% of pediatric rhabdomyosarcome occurs in the head and neck [1]. Which is the opposite for adults where the head and neck sarcoma represented only 1% of all head and neck cancers [4].

The commun type in adults is the pleomorphic rhabdomyosarcoma but it is very rare in the head and neck region. In addition, this tumor has a higher rate of recurrence and metastasis in adults [3].

The clinical presentation of the head and neck rhabdomyosarcoma depends on its location, the patients usually present a rapidly growing, painless, swelling [4,5]. The radiological examination that can be used to identify the extension of the tumor in this region are the cranio-facial CT scan or an MRI.

The diagnosis of the pleomorphic RMS is made by the histopathologic study, that usually shows a large, atypical pleomorphic tumor cells with abundant eosinophilic cytoplasm which can be irregular and hyperchromic, they also can have large or multiple nuclei. The diagnosis is confirmed by the immunohistochemical tests, the tumor cells express desmin, actin, myogenin, Myoglobin, Vimentin and MyoD1 [3-5].

Differential diagnosis of a head and neck mass should always be considered to avoid any misdiagnosis and mistreatment of the patients. Other malignant tumors with rhabdomyosarcomatous differentiation need to be considered on the differential diagnosis. Such as neuroblastoma, lymphoma, melanoma, fibrosarcoma, granular cell myoblastoma, rhabdomyoma, Ewing's sarcoma, chordocarcinoma and sarcomatoid carcinoma etc [4].

For our patient, the immunohistochemistry test confirmed the diagnosis by the presence of desmin, actin and myogenin, which are highly sensitive and specific markers for RMSs

RMSs are locally aggressive and they can spread regionally, or distantly by both lymphatic and hematological routes. The commonest sites of metastatic spread are lung, liver, lymph node and bone [6]. Therefore, during follow up, Computerized tomography (CT) examinations of the abdomen and thorax are essential to search any metastases or lymphadenopathy in the neck.

Several prognostic indicator are used to evaluat the prognosis of the RMS, which include, age, tumor location, size, histologic subtype, and the presence or absence of regional nodes or distant metastases.The age seems to be the most important prognosis factor, studies showed that Children treated for RMS had a significantly better outcome than adults regardless the subtype of the RMS [6].

The treatment of patients with malignant mesenchymal tumors of the head and neck region is multidisciplinary and multimodal includes radical surgery with postoperative radiotherapy and or chemotherapy.

The surgical execison of the tumor should be radical to prevent any local reccurence. The analysis of the specimen and the assessment of the extent of the tumor allow to establish the stage of the tumor according to the Intergroup Rhabdomyosarcoma Study (IRS) [7].

Radiotherapy is also an integral part of the treatment. The irradiation targets the tumor and the lymph nodes. It is not systematic, it is used as adjuvant therapy for small volume residual disease after surgery [8].

chemotherapy contributes to improved survival and reduced frequency of recurrence, also reduces the indications of extensive surgical excisions.

However, in high-risk metastatic forms, the treatment approach can be more conservative, partial surgical excision or non-surgical therapies such as radiotherapy and or chemotherapy [8].

In our case, the clinical and radiological présentation of the tumor didn't give us a diagnosis orientation. That is why we proceeded by an intra-operative frozen section that showed the présence of signs of malignancy. So we proceeded by the resection of the tumor with wide margins

The histological test of the surgical specimen confirmed the diagnosis of pleomorphic rhabdomyosarcoma and the total excision of the tumor. However, considering the malignancy and the metastatic potential of the tumor and the age of the patient, he was referred to the oncology center for a complement of radiotherapy.

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