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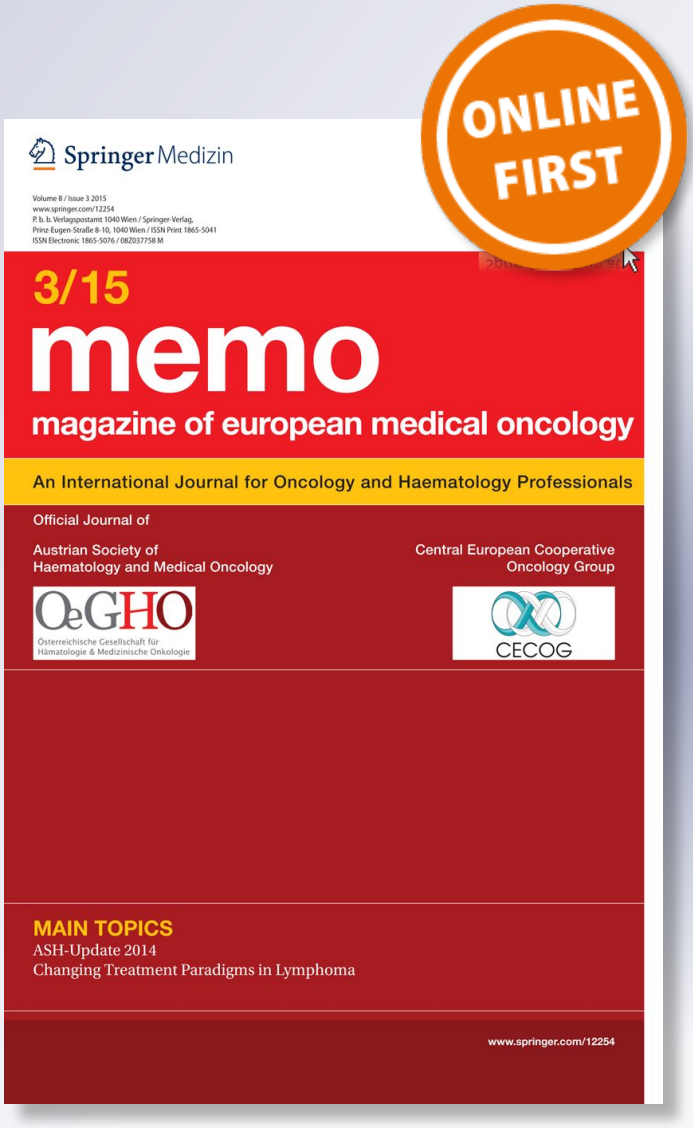

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Orbito-maxillofacial mesenchymal chondrosarcoma with intracranial invasion and lung metastasis

Case report and review of the literature

Ilson Sepúlveda · Max Schorwer · Enrique Platin · Pablo Mucientes · Álvaro Compan · Joaquin Ulloa · Jaime Pinto · Nestor Quitral

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Summary We report on a patient who presented with swelling to the left side of the face accompanied by exophthalmos. Imaging studies revealed a large expansive heterogeneous mass encroaching on the sinonasal region. Following intravenous contrast administration revealed a lesion with moderate enhancement and involvement of the ipsilateral orbital cavity with intracranial invasion. The patient underwent tumor resection and a biopsy confirmed the presence of a mesenchymal chondrosarcoma. Additional treatment included radiation therapy and the patient is currently receiving palliative and supportive care for advanced metastatic lung disease.

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Keywords Tumor · Head · Neck · Magnetic resonance imaging (MRI) · Computed tomography (CT)

Introduction

Chondrosarcomas (CHS) are uncommon malignancies that usually arise from cartilaginous, bony, or soft tissues. They mainly affect males between the third and fourth decades of life. In the head and neck these tumors are locally aggressive, but metastasis to the lymph nodes of the neck is rare. Hematogenous metastasis occurs more frequently to the lung. Imaging studies are essential for preoperative staging and surgical planning. The treatment of choice is complete surgical resection.

Case report

A 30-year old female without a significant medical history presented to the ophthalmology clinic complaining of progressive left exophthalmos of 4 months duration. No pain or neurologic deficits were present. A computed tomography (CT) scan was performed, revealing an aggressive, expansive, and destructive heterodense solid mass on the left side of the face. The mass displayed irregular contours with central hypodense areas following intravenous contrast administration, suggestive of necrosis. The mass extended from the maxillary region, invading the ipsilateral orbital and nasal cavities, ethmoid sinus, masticatory space, and the supra- and infratemporal fossa (Fig. 1). There was no evidence of cervical lymphadenopathies and the CT of the thorax was normal.

In addition, an MRI was performed, showing a partially delimited expansive process. T1-weighted MRI showed a mildly hypointense solid mass centered in

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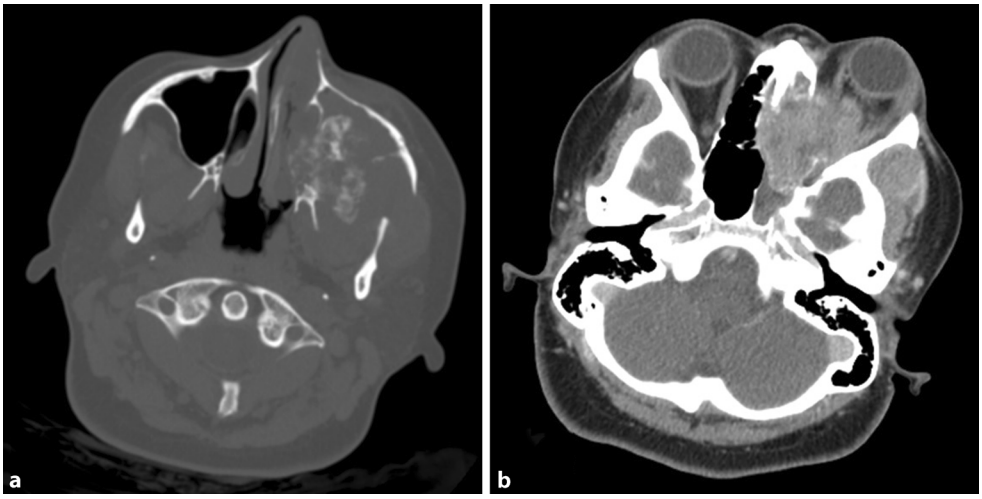

Fig. 1 Computed tomography (CT) showing a primary sinonsal expansive process with orbital cavity and temporal fossa involvement. **a** CT bone window axial view. **b** CT soft tissues window after intravenous contrast injection

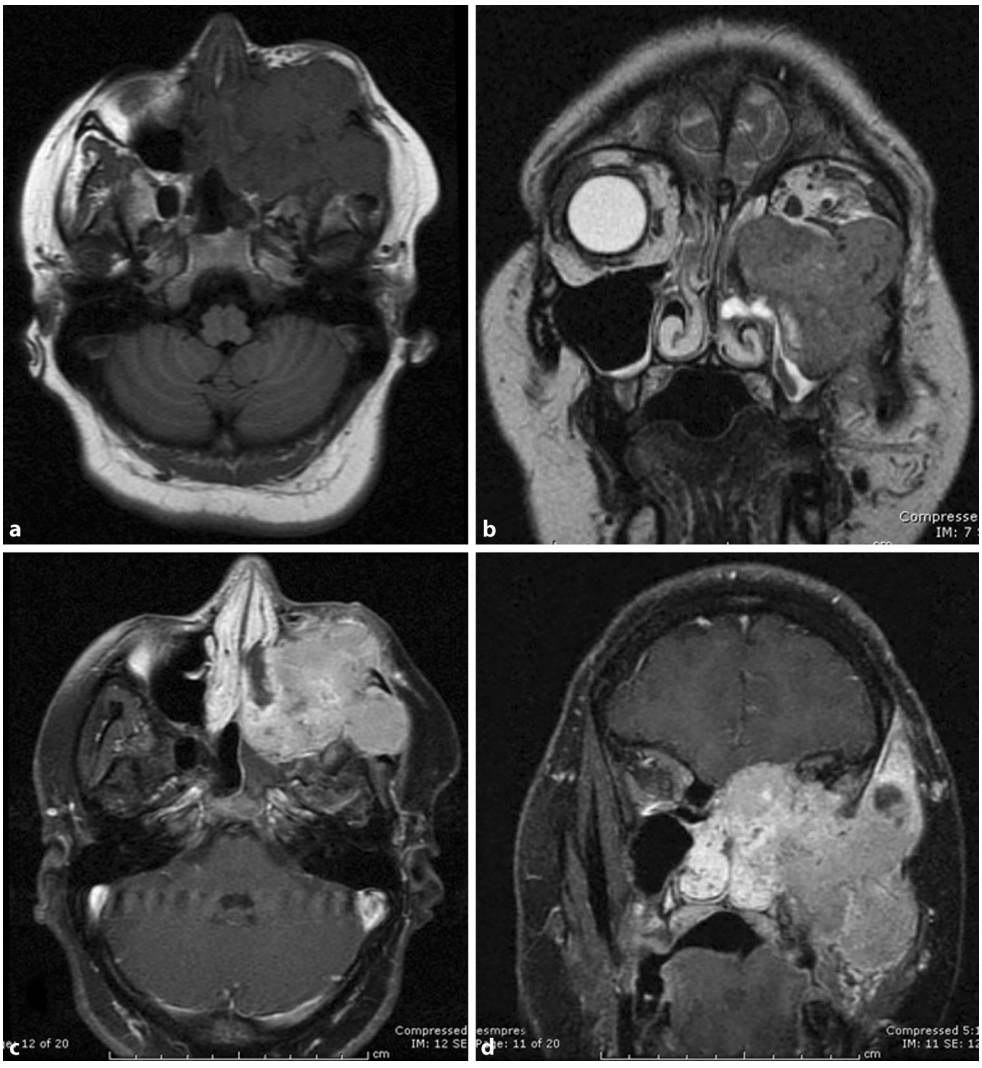

Fig. 2 Magnetic resonance imaging (MRI). Large solid expansive left orbito-maxillofacial process with intracranial invasion and moderate enhancement following intravenous injection of paramagnetic contrast medium. **a** T1 axial view. **b** T2 coronal view. **c** T1 fat-saturated gadolinium axial view. **d** T1 fat-saturated gadolinium coronal view

the left maxillary region invading the left side of the ethmoidal sinus and nasal cavity. T2-weighted MRI showed a mildly hyperintense mass invading the left orbit with lateral displacement of the medial and inferior rectus muscles and globe. Intracranial invasion with left cavernous sinus involvement and heterogeneous enhancement of the mass was seen in the post-contrast fat-saturated images (Fig. 2).

A biopsy was performed under general anesthesia revealing a CHS. The Head and Neck Tumor Board

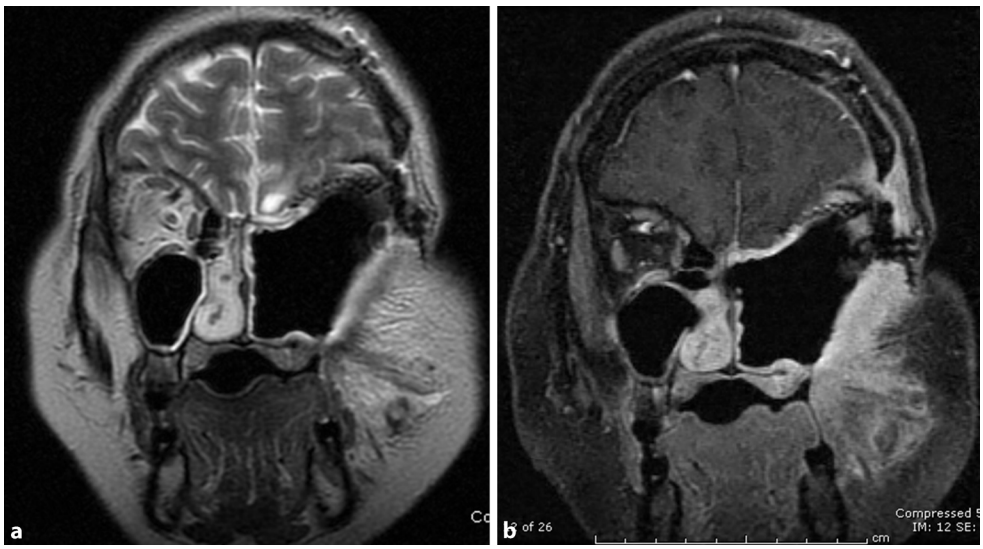

Fig. 3 Magnetic resonance imaging (MRI). Control study after surgical treatment of the primary tumor. **a** T2 coronal view. **b** T1 fat-saturated gadolinium coronal view

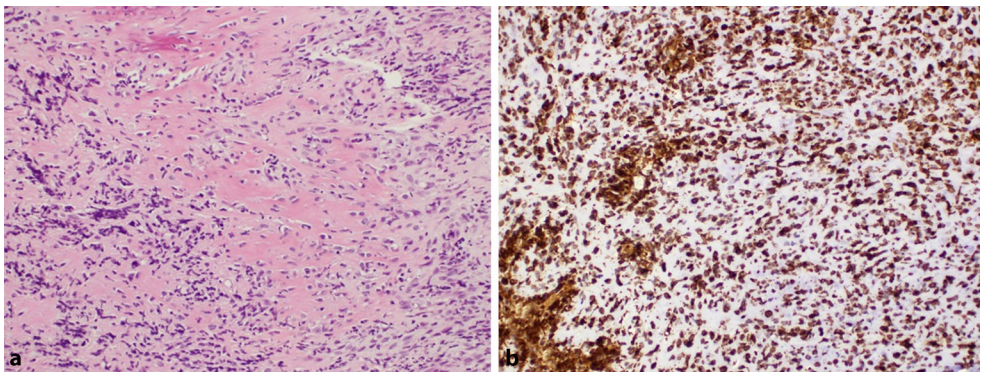

Fig. 4 Immunohistochemistry study. **a** Hematoxylin and eosin stain. **b** Positive BCL-2 stain. Magnification x 20

(HNTB) reviewed the case, where it was staged as a T4N0M0 orbito-maxillofacial tumor. The HNTB recommended surgical excision in a multidisciplinary approach as the best treatment option due to the size of the mass (Fig. 3). The postsurgical biopsy confirmed the presence of a mesenchymal chondrosarcoma with infiltration of adjacent structures. Immunohistochemistry tests were positive for BCL-2 and negative for myogenin, CD99, CD34, S-100, desmin, vimentin, actin, chromogranin, and synaptophysin (Fig. 4).

Follow-up by the HNTB resulted in the recommendation of adjuvant radiation therapy. Conventional radiotherapy was performed with linear accelerator, with a total dose of 60 Gy administered in 2-Gy doses during a 6-week period (Fig. 5). At the end of the treatment, neck and chest CT examinations were performed, revealing the presence of nodules in the left lung suggestive of metastatic disease (Fig. 6). The patient was referred to the pain control and palliative care program.

Discussion

Sarcomas are very rare among head and neck neoplasms. They arise from transformed cells of mesenchymal origin, representing only 1% of all primary tumors. Current classification schemes attempt to group sarcomas into subtypes that are useful for determining prognosis and formulating treatment strategies, where only 20% have bony or cartilaginous origin [1]. CHS are rare malignancies [2] most commonly found in the pelvic bones, proximal and distal femur, proximal humerus, and the ribs [3, 4]. CHS in the head and neck are uncommon, accounting for less than 12% of all cases and only 0.1% of head and neck malignancies [5, 6]. They arise from bone, in which the tumor cells originate from chondroid and not osteoid bone [4]. They are characterized as slow growing in adult males with mean ages ranging from 35 to 45 years [6]. Patients with Maffucci syndrome and Ollier disease have a 25–30% risk of developing CHS [3].

The most common locations of CHS in the head and neck include the nasal cavity, maxilla, skull base, paranasal sinuses, and mandible [4]. The clinical finding is usually painless swelling, expansion of cortical plates, nasal discharge, facial paralysis, and bleeding [7]. When the tumor is growing, the patient may have a headache and neurologic problems, depending on the location [3].

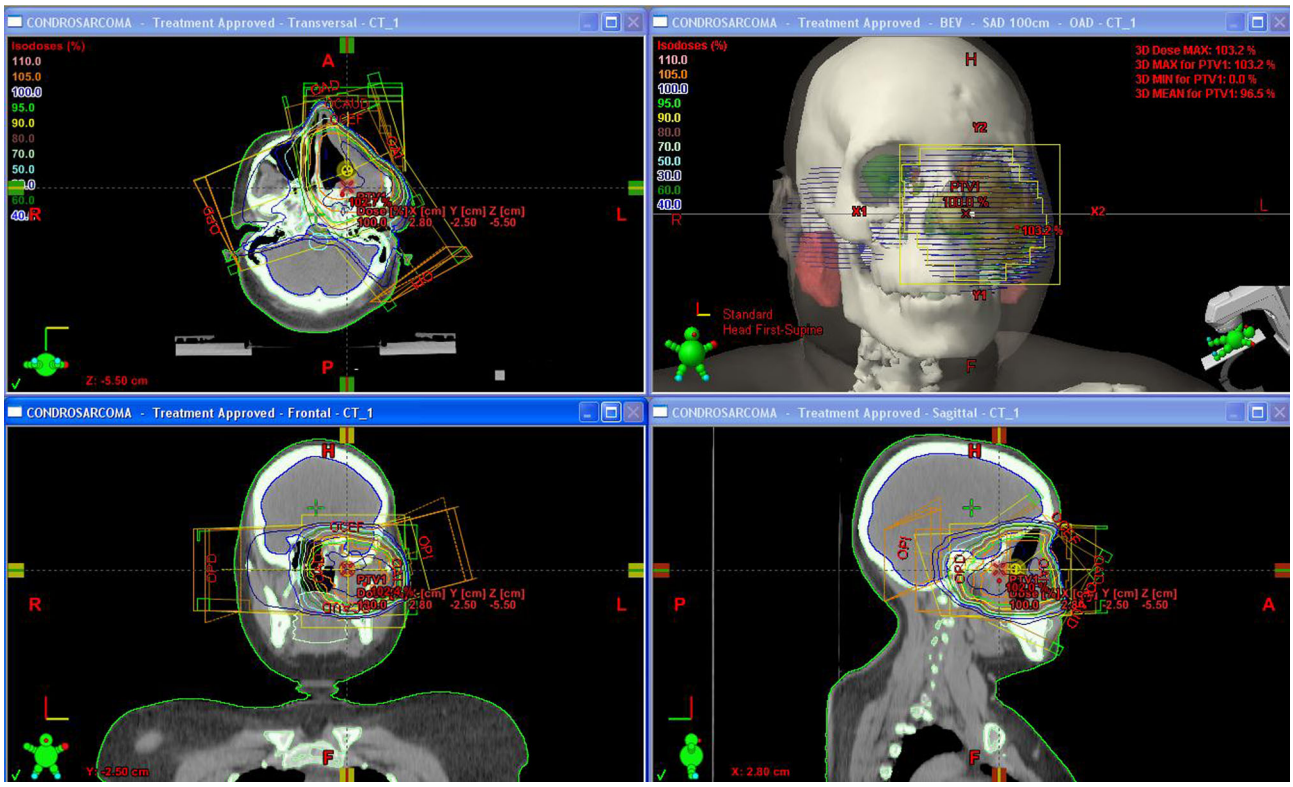

Fig. 5 Three-dimensional conformal radiotherapy plan

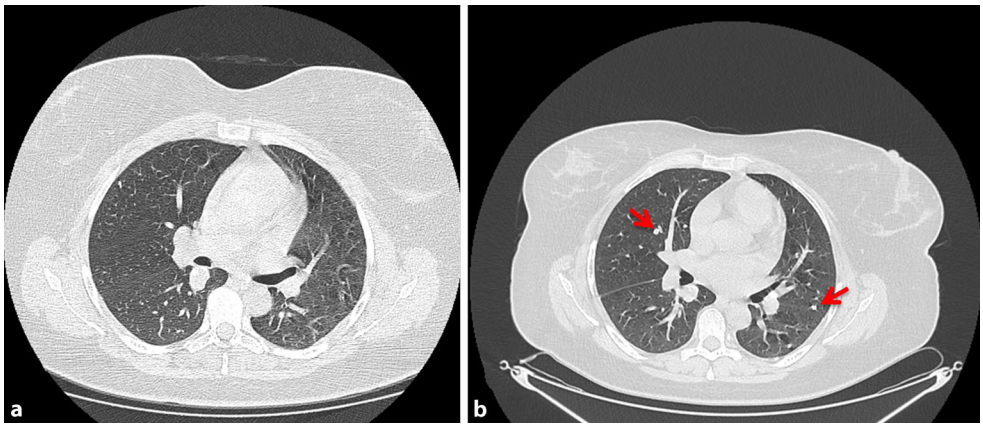

Fig. 6 Comparative CT scans. **a** September 2015 showing no evidence of pulmonary nodules. **b** July 2016 showing multiple metastatic nodules in both lungs (*red arrows*)

The pathologic types of this tumor include mesenchymal myxoid and low grade. CHS are composed of lobulated cartilage without osteoid tissue, round and oval cells in lacunae with enlarged nuclei. Atypical spindle cells, myxoid degeneration, and calcification or ossification of the matrix can be seen [3, 5, 6, 17]. The classification of histologic malignancy for CHS was proposed by Evans et al. in 1977, and it is graded I through III depending on the mitotic rate, cell density, and nuclei size [8]. The histologic grade is a consistent predictor of prognosis and its importance is illustrated in the American Join Committee on Cancer (AJCC) staging system for sarcomas [9].

Since sarcomas are frequently associated with soft tissue invasion and bone destruction, CT and MRI are essential for preoperative staging and surgical planning in patients with these tumors. CT scan findings include single or multiple hypodense areas, bone sclerosis, endosteal scalloping, and tumor calcifications. In late-stage disease, the primary lesion may penetrate the cortical plate, causing cortical expansion, destruction, and extension into adjacent soft tissues [5–7, 10, 11]. In MRI, in general, the CHS have high signal intensity on T2-weighted images. Bone deposits in the tumor appear with decreased signal intensity in all sequences [4, 10, 11, 15].

The management of the sarcomas is not always applicable to the head and neck, due to complex anatomy and the difficulty of obtaining wide surgical margins. This determines the higher recurrence rates and worse disease-specific survival rates in comparison with sarcomas arising at other sites [12]. Complete resection is the most effective primary treatment and may include soft and hard tissues [3, 5, 13, 19]. Positive margins and residual tumor is a powerful predictor of local recurrence. Neck dissection is not indicated due to the low incidence of cervical node metastasis [7, 14]. Adjuvant radiation therapy is recommended after surgery with positive margins and regional lymph node dissection for nodal metastases. This, however, does not improve the survival rates, because patients succumb to distant metastases [4, 11, 19].

Proton beam radiotherapy (PBRT) has been proposed to treat targets near critical structures, such as the skull base, to lower normal tissue exposure without compromising target coverage, thereby improving the therapeutic ratio. Multiple publications have shown that PBRT results in favorable clinical outcomes compared to photon therapy for tumors of the skull base and paranasal sinuses [20, 21].

Chemotherapy is used for palliation or, in cases of aggressive behavior or potential metastases, as adjuvant therapy for maximizing local control and treating the potential of micrometastatic disease [2, 6, 7, 14, 19].

The rate of local and distant metastases is round 5%. Metastases have been reported more frequently in the lung [6, 10]. The 5- and 10-year survival rates for head and neck CHS ranges from 50 to 60%, according to the histologic grade of malignancy and surgical margins. Strict clinical and imaging (CT) follow-up is required [3, 5, 10, 16, 18].

Conclusion

Chondrosarcomas are a rare entity arising from cells of mesenchymal origin. Imaging studies play an important role in treatment planning, providing valuable information about the growing pattern and involvement of critical anatomic organs. The treatment of choice is surgical resection, combined with adjuvant radiation therapy in some cases.

Conflict of interest I. Sepúlveda, M. Schorwer, E. Platin, P. Mucientes, Á. Compan, J. Ulloa, J. Pinto, and N. Quitral declare that they have no competing interests.

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