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Case report

Chondrosarcoma of the parapharyngeal space: Case report and literature review of an extremely rare location ☆☆☆

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ABSTRACT

Chondrosarcoma of the head and neck region is a rare disease, representing approximately 0.1% of all head and neck neoplasms. Parapharyngeal location is extremely rare and low-grade ones are even rarer. Surgery alone or followed by adjuvant radiotherapy is the treatment of choice. In this article, we report a case of a 67-year-old male with low-grade parapharyngeal chondrosarcoma who presented with a 3-month history of dysphagia. A cervical magnetic resonance imaging was performed that shows a well-defined mass located at the right parapharyngeal space, causing medial deviation of the mucosal space. Surgical resection of the tumor without neck dissection followed by adjuvant radiotherapy was undertaken with a favorable response. The purpose of this article is to add our case to the limited literature for good management of parapharyngeal chondrosarcomas.

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Background

The parapharyngeal space, also known as the prestyloid parapharyngeal space, is located in the deep part of the head and neck.

Chondrosarcomas of the head and neck region are rare. There are usually slow-growing lesions. Chondrosarcomas of the parapharyngeal space are limited in number and among them no low-grade chondrosarcomas. We report in this article a case of low-grade parapharyngeal chondrosarcoma treated using simple excision without dissection of

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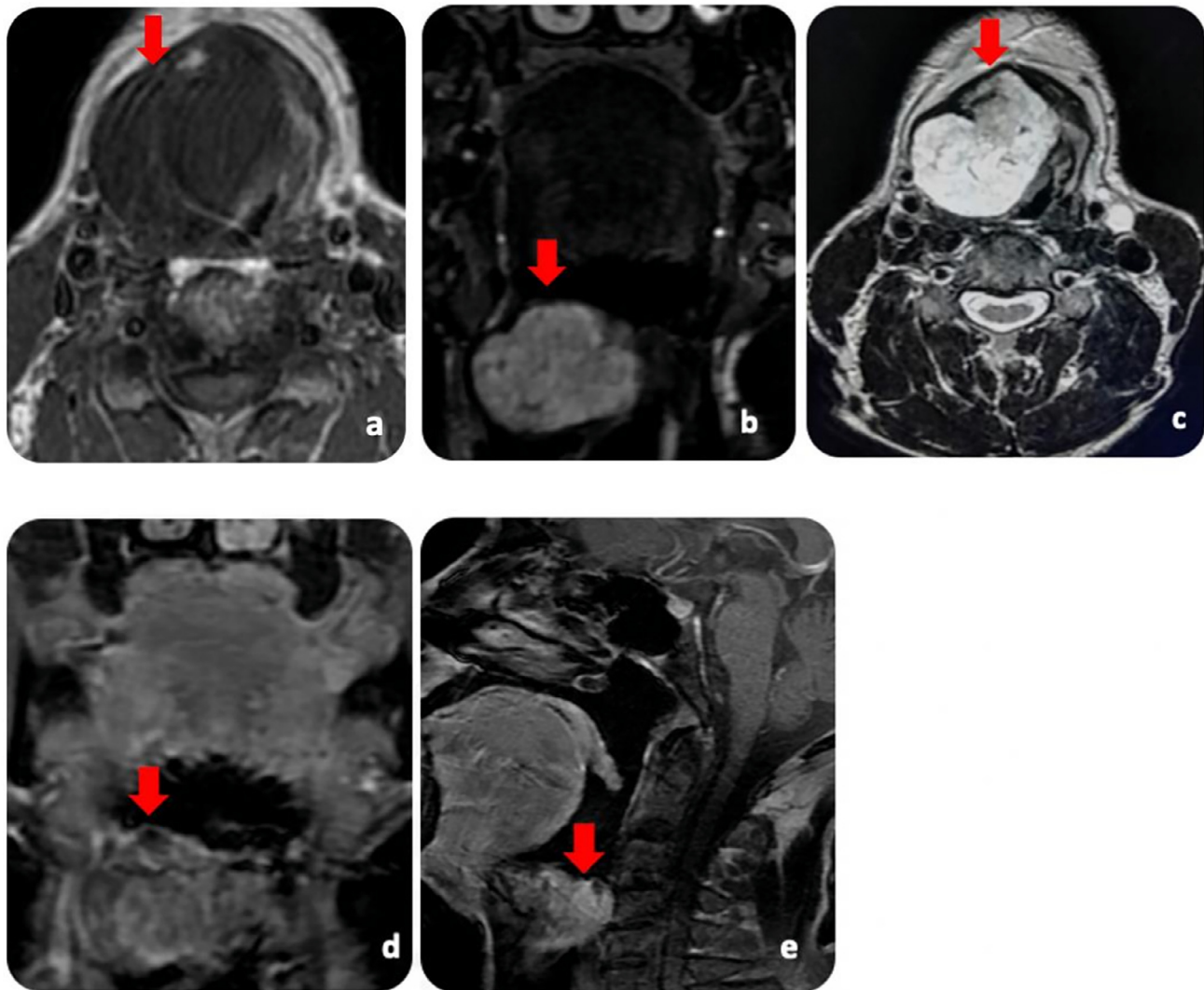


Fig. 1 – Preoperative MRI of the lesion showing: A well-defined mass located at the right parapharyngeal space, causing medial deviation of the mucosal space (Red arrow).

- Low signal intensity of tumor in T1W image (A).
- High signal intensity of tumor in T2W images: coronal (B) and axial (C) plans.
- Post contrast weighted images in Coronal (D) and sagittal (E) plans revealing an heterogeneous enhancement of the tumor. (Color version of figure is available online.)

the neck followed by radiation therapy with a favorable response.

Case presentation

A 67-year-old nonsmoking man presented to our Hospital for mild dysphagia to solid. No family history of head or neck tumors or vascular abnormalities.

The clinical examination found the patient in good general condition. Performance status was 0-1, with a right cervical swelling of 3 cm. No inflammatory signs or pain on palpation.

Cervical ultrasonography was indicated that shows a mass of the upper cervical part of the carotid axis, well-

circumscribed, hypoechoic, with macrocalcification, poorly vascularized evoking probably a pleomorphic adenoma of an accessory salivary gland.

To complete the investigation, a MRI was performed that shows an encapsulated, well-defined mass located at the right parapharyngeal space, causing medial deviation of the mucosal space. No signs of invasion of adjacent structures or intracranial extension were observed (Fig. 1).

The patient underwent surgical treatment with complete removal of the lesion.

The pathologic examination concluded to well-differentiated cartilaginous neoplasm of slightly atypical low cell density confirming the diagnosis of low-grade chondrosarcoma.

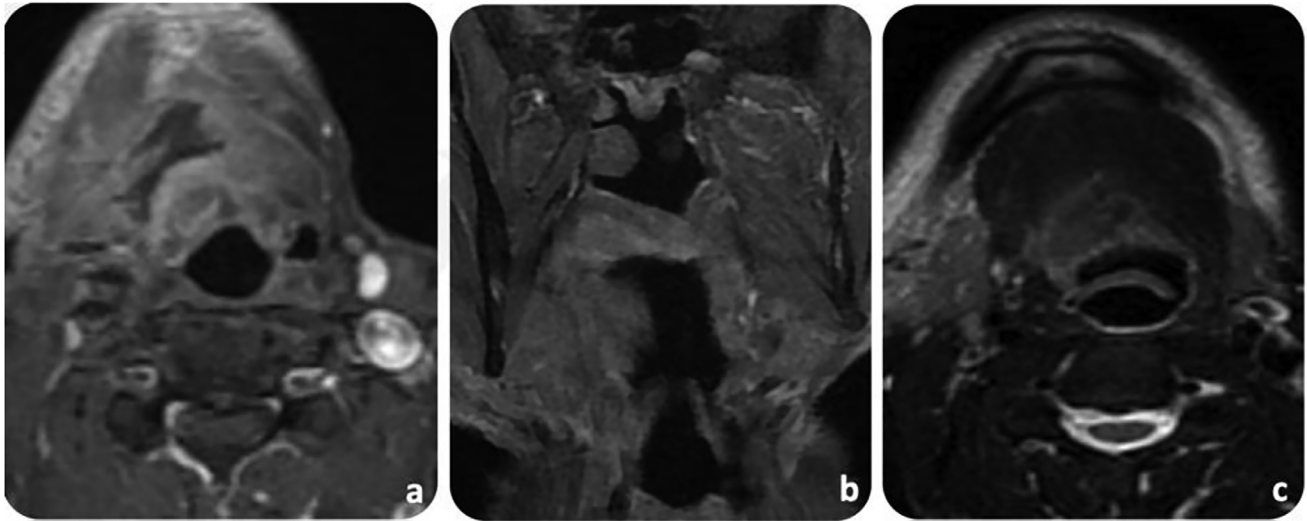


Fig. 2 – MRI weighted post contrast images in Axial (A) and coronal (B) plans showing a complete removal of the tumor.

Post-operative MRI had shown the persistence of some inflammatory changes in the right parapharyngeal space. (Fig. 2).

The patient had received adjuvant external radiotherapy at a dose of 60 Gy, 2 Gy/fraction, 5 fractions/week without side effects.

During the 18 -months follow-up, no residual tumor was observed.

Discussion

Chondrosarcoma is a malignant tumor that represents about 20% of malignant bone tumors. It is characterized by its slow growth and hyaline cartilage differentiation.

Its localization in the head and neck is rare, representing less than 12% of all HSC cases [2] and only 0.1% of malignant tumors of the head and neck [1–3].

Most often, the anterior part of the maxilla, the sino-nasal region, and the mandible are affected [4], the parapharyngeal space location is extremely rare. As a result, very few clinical series concerning the clinicopathological characteristics neck have been published in the literature.

Chondrosarcomas of the parapharyngeal space are rare and among them, there are no low-grade chondrosarcomas. Only a few cases had been reported.

Three of them are tumors that reach the parapharyngeal space from adjacent structures and were all high-grade tumors [5].

Our case differs from the others in the sense that it is low-grade chondrosarcoma of this space.

Chondrosarcoma can present with a variety of clinical symptoms but painless cervical swelling remains the most common symptom [4].

MRI is the best modality to appreciate tumor size, extent, and the relationship to adjacent structures.

The cornerstone of head and neck chondrosarcoma treatment is surgical resection

The type of surgery depends on the histological grade, extension, and location of the tumor. The key point for local control is to obtain adequate surgical margins [1,6].

Opinions are divided on the radiosensitivity of head and neck chondrosarcoma. Since it is a slow-growing tumor with a small fraction of dividing cells, it is considered “relatively” radioresistant.

Radiotherapy treatment is mainly offered as adjuvant therapy, in cases of highly probable residual disease after surgery [7], and palliation in patients with unresectable lesions or unsuitable for major surgery [8].

In low-grade chondrosarcoma with negative surgical margins, adjuvant radiotherapy is not indicated. In high-grade chondrosarcomas, adjuvant radiotherapy is appropriate because of the aggressiveness of the tumor [9].

Our case was low-grade chondrosarcoma and for this reason, wide excision without neck dissection was enough for surgical procedure. On the other hand, patients with high-grade chondrosarcomas usually need adjuvant therapy [10,11].

Generally, the disease prognosis is reported to be poor with 5-year survivals ranging from 30% to 50% [12]. Distant metastases occur in only 7–18% of cases. However, high-grade tumors can metastasize 71% preferentially in the lung and bone.

Surgical treatment of head and neck tumors improves the prognosis.

Other factors influencing the prognosis are the tumor size and stage, the histopathological grade of the lesion (poor prognosis for high-grade tumors), and the localization.

Long-term follow-up is recommended because tumors may lead to local recurrence or even metastasis several years after initial diagnosis [1,6].

In conclusion, the purpose of this report is to add our case to the limited literature and make better-informed choices to the management of chondrosarcomas of the parapharyngeal space.

Authors' contributions

All the authors participated in the writing of the article and the bibliographical research concerning the case described. They also declare having read and approved the final version of the manuscript.

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