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Alveolar rhabdomyosarcoma of the mandible in an adult invading the parotid gland: A rare case report

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ABSTRACT

INTRODUCTION: Rhabdomyosarcoma (RMS) is a malignant mesenchymal neoplasm uncommon in adults.
PRESENTATION OF CASE: We report the case of an alveolar rhabdomyosarcoma in a rare location and in an adult. It concerns a 46-year-old woman with a mandibular location invading the parotid gland. The Immunohistochemical analysis revealed intense positivity for myogenin and desmin, favoring the diagnosis of alveolar rhabdomyosarcoma. The patient had a right hemi-mandibulectomy with parotidectomy and received chemotherapy with radiotherapy. The evolution was good up to 2 years postoperatively then the patient relapsed with a recurrence of tumour rapidly progressing and metastases in the cervical spine.
DISCUSSION: RMS is an aggressive but rare disease that is one of the most common malignant head and neck tumors in children. The predilection sites of adult rhabdomyosarcoma are the extremities. Current treatment includes a combination of ablative surgery, chemotherapy, and radiation therapy.
CONCLUSION: The prognosis of head and neck rhabdomyosarcomas in adults remains very poor given the often initially advanced stage of the disease and the high metastatic potential.

1. Introduction

Rhabdomyosarcoma (RMS) is a skeletal muscle tumour divided into three main groups: embryonal, alveolar and pleomorphic [1].

Alveolar rhabdomyosarcoma predominates in an older age group (10–25 years) than embryonic rhabdomyosarcoma. It also appears in the head and neck, including the parameningeal region [2].

Later studies have shown that in older patients, these lesions occur mainly in the third and fourth decades and show a striking predilection for the head and neck region [3].

Treatment options include surgery, radiation therapy, and chemotherapy.

The work has been reported in line with the SCARE criteria [4].

2. Presentation of case

A 46-year-old woman presented to the Otolaryngology Department, for evaluation of a huge mandibular angle mass extending to the parotid region, increasing in size progressively over 6 months. The patient reported chewing discomfort with limited mouth opening, but denied any problems with speech or swallowing, without peripheral facial paralysis facial.

Physical examination found a mass of the right hemiface, voluminous, measuring about 10 × 8 × 7 cm, fixed in relation to the deep plane, painless (Fig. 1) with V3 right paraesthesia.

Intra-oral examination revealed vestibular bulging, without trismus or cervical lymphadenopathy.

A panoramic x-ray showed an ill-defined radiolucent lesion, with osteolysis on the right mandibular branch extending to the lower edge of the mandible body (Fig. 2).

Computed tomography (CT) scan demonstrated the presence of a seat tissue density process enhanced heterogeneously on the angle of the right mandible, with osteolysis. It extends to the region of the parotid gland, with erosion of the upper edge of the mandibular canal (Fig. 3).

A positron emission tomography (PET) scan showed the hypermetabolic mass of the soft tissues of the right face (44 × 38 mm). without hypermetabolic right cervical lymphadenopathy or hypermetabolic focus identifiable elsewhere.

Abbreviations: RMS, rhabdomyosarcoma; CT, Computed tomography.

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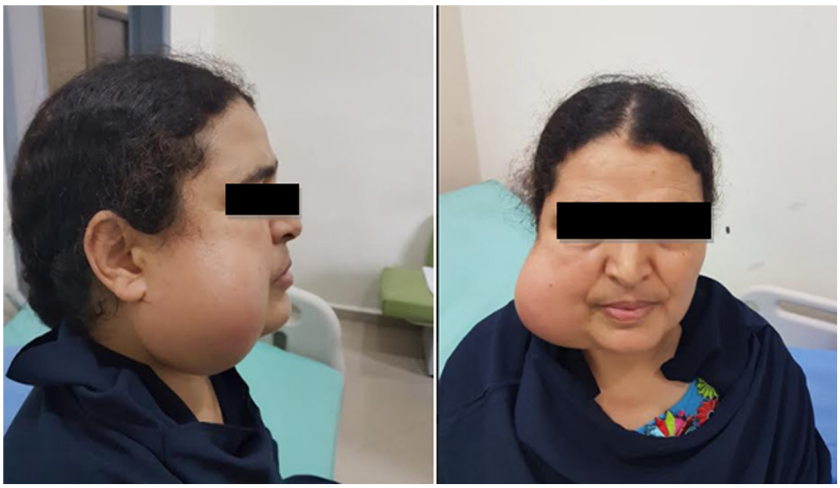

Fig. 1. Frontal (A), and profile (B) views demonstrating the right-sided facial mass.

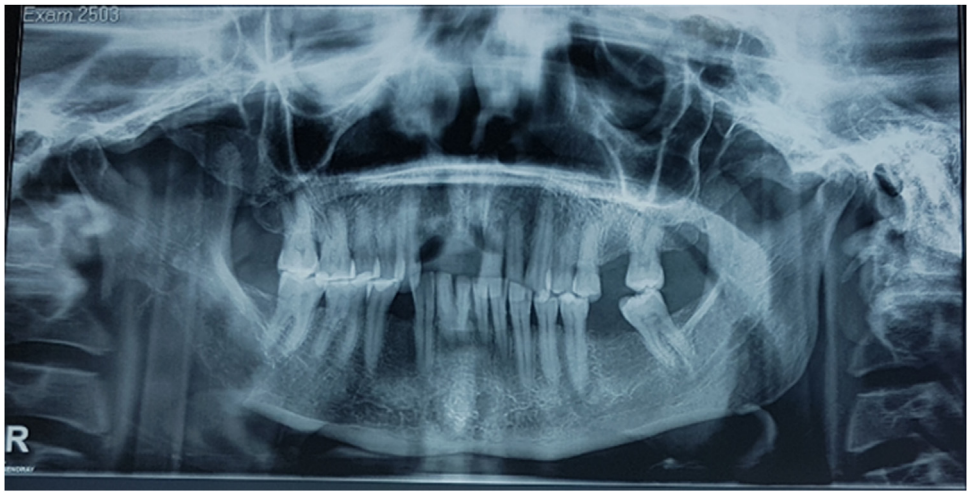

Fig. 2. Ill-defined radiolucency of the right mandibular ramus with cortical destruction are apparent on panoramic radiograph.

Histological examination revealed large round cells with hyperchromatic nuclei and an eosinophilic cytoplasm (Fig. 4A); further immunohistochemical studies were conducted to further classify this lesion. Neoplastic cells were positive for myogenin (Fig. 4B) and desmin (Fig. 4C).

Based on the immunohistochemical results, a diagnosis of alveolar RMS was made.

The staging of RMS before treatment was determined by the location of the tumor (head and neck), the size of the tumor (> 5 cm in diameter), the nodal involvement (without affecting the regional lymph nodes in our patient) and distant metastases (none).

Based on these criteria, the patient underwent local surgical resection, she underwent a right hemi-mandibulectomy with parotidectomy.

The final resection sample consisted of part of the mandible and associated teeth measuring 13 × 12 × 8 cm and extending to the right parotid gland (Fig. 5A–B).

The histological analysis of the resection sample was consistent with the results observed during the initial biopsy (Fig. 4A–C). The histological margins were free and the resection complete.

The patient received postoperative chemotherapy and radiotherapy.

The evolution was good for the first 2 years. The patient presented no symptoms and her condition progressed well. The control CT scan showed no residual lesions or signs of recurrence.

After 2 years, the patient unfortunately experienced a recurrence with a rapidly progressing tumour and metastases in the cervical spine that did not respond to rescue chemotherapy. She received additional palliative radiotherapy to manage the local symptoms

3. Discussion

Rhabdomyosarcoma (RMS) is highly malignant tumors that originates from immature cells that are destined to differentiate into striated skeletal muscle [5].

The histological aspect of alveolar RMS is characterized under the microscope, by round or oval tumor cells separated into nests by partitions of connective tissue [6].

The only really reliable way to reach the diagnosis is the immunohistochemical demonstration of the positivity of Myogenin and desmin in a significant proportion of tumor cells [3].

Myogenin is the most sensitive and specific marker of rhabdomyosarcoma, it is expressed in a particularly strong and diffuses way in the alveolar type [7].

Desmin is an intermediate filament that is a specific indicator for muscle differentiation, but it is present in both smooth and striated muscle [8].

The true etiology of RMS is unclear.

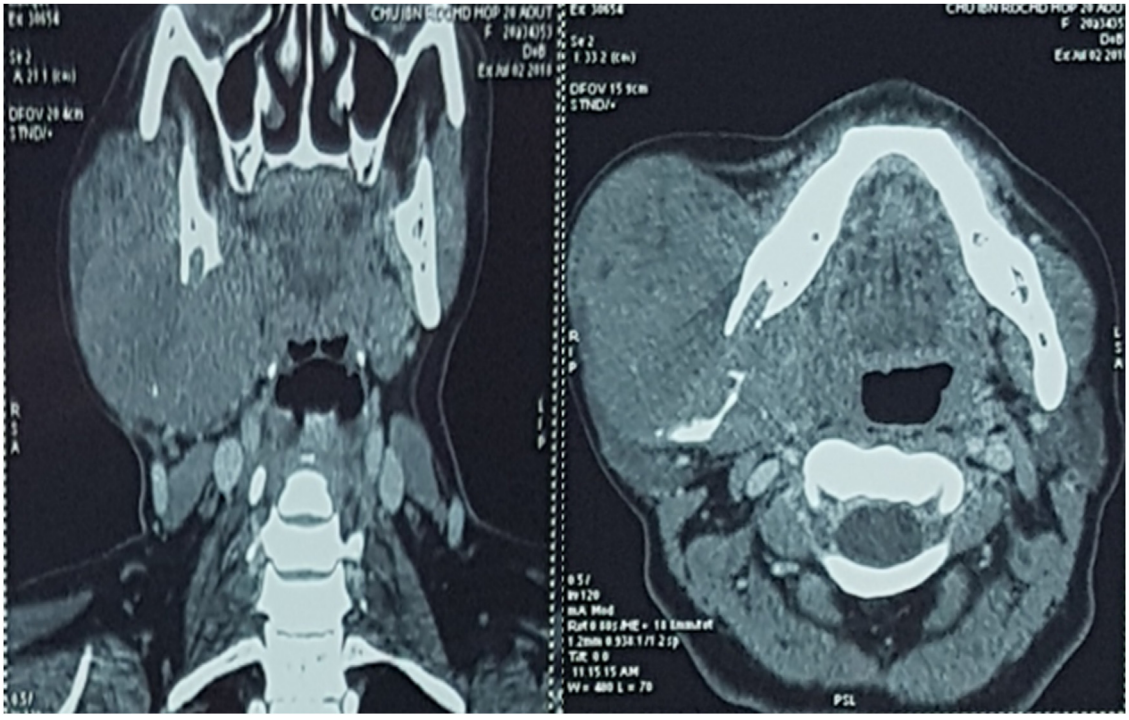

Fig. 3. CTscan has demonstrated the presence of a solid heterogeneous mass of 5.5 cm centered on the right mandibular branch, with osteolysis,extending to the region.

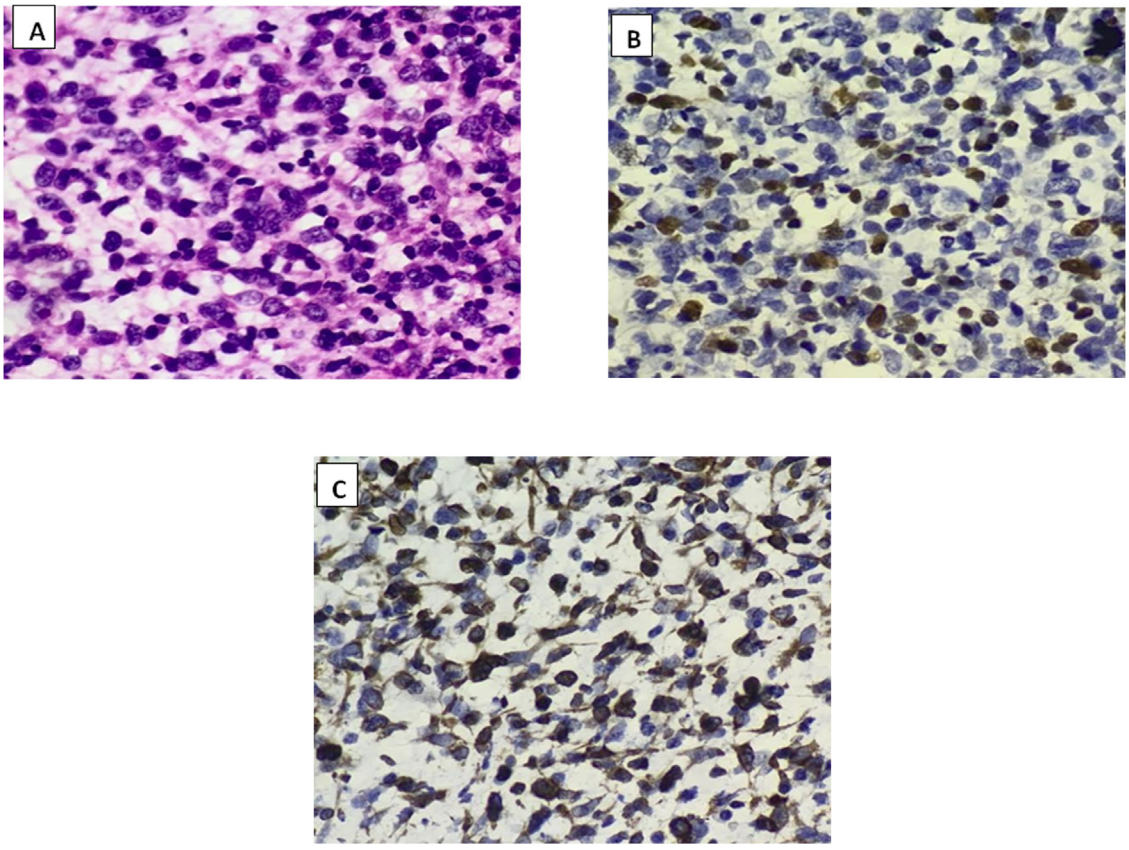

Fig. 4. A. Alveolar rhabdomyosarcoma showing a giant multinucleated tumor cell with hyperchromatic nuclei and eosinophilic cytoplasm. B. The lesional cells stained positive for myogenin. C. The lesional cells stained positive for desmin.

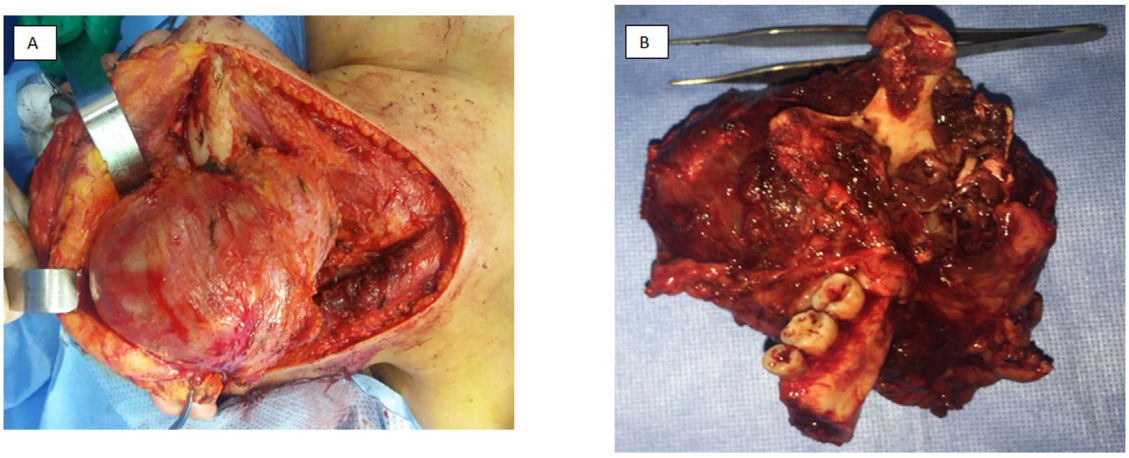

Fig. 5. A. Intraoperative photo showing a mandibular angle mass arriving at the parotid gland. B. Operative part of a hemimandibulectomy with an immediate macroscopic postoperative aspect of an alveolar rhabdomyosarcoma.

Symptoms of RMS are variable, depending on the site of initial presentation, the extent of the tumor, and the presence or absence of distant metastases and lymph node involvement [9].

The radiological characteristics of the tumor are not specific.

CT scanning is useful to assess bone erosion. Magnetic resonance imaging gives a better definition of the mass and its invasion of adjacent structures.

Common sites of metastasis reported by conventional investigations are the lungs, skeletal system, lymph nodes, and brain, with hematogenesis being the common route of metastasis [10].

The standard treatment for adults with localized soft tissue sarcoma is based on surgery, often complemented by radiotherapy. To date, some benefit resulting from adjuvant chemotherapy has been demonstrated in adults with soft tissue sarcoma [11].

Radiotherapy is only reserved for patients who develop a recurrence after the end of the initial treatment. Our patient received radiation therapy due to the invasion of the parotid gland.

The prognosis depends on histology, primary site, size of the tumor and extent of disease; in which tumors are classified as having either localized, regional, or distant extension. The RMS with alveolar histology had substantially worse survival as compared with those with embryonal histology [11].

4. Conclusion

RMS is an aggressive but rare disease that is one of the most common malignant head and neck tumors in children. The predilection sites of adult rhabdomyosarcoma are the extremities.

Immunohistochemical staining with muscle-related antigens (desmin, myogenin) is necessary to confirm the myogenic nature and confirm sarcomeric differentiation.

Current treatment includes a combination of ablative surgery, chemotherapy, and radiation therapy.

The prognosis is worse in adults, due in part to the aggressive biological behavior of RMS.

Conflicts of interest

No conflicts of interest.

Funding

Personal sources.

Ethical approval

Ethical approval has been exempted by my institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

All the authors worked in coordination to ensure the best management of the disease from diagnosis to surgery with close postoperative monitoring. The authors wrote this article together.

Registration of research studies

1. Name of the registry: Research Registry
2. Unique identifying number or registration ID: researchregistry5954
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): https://www.researchregistry.com/browse-the-registry#home/

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References

[1] I. Sultan, I. Qaddoumi, S. Yaser, C. Rodriguez-Galindo, A. Ferrari, Comparing adult and pediatric rhabdomyosarcoma in the surveillance, epidemiology and

end results program, 1973 to 2005: an analysis of 2,600 patients, J. Clin. Oncol. 27 (20) (2009) 3391–3397.
[2] T. Yasuda, K.D. Perry, M. Nelson, et al., Alveolar rhabdomyosarcoma of the head and neck region in older adults: genetic characterization and a review of the literature, Hum. Pathol. 40 (2009) 341–348.
[3] Christopher D.M. Fletcher, Recently characterized soft tissue tumors that bring biologic insight, Mod. Pathol. 27 (Suppl 1) (2014) S98–S112.
[4] R.A. Agha, M.R. Borrelli, R. Farwana, K. Koshy, A. Fowler, D.P. Orgill, For the SCARE Group, The SCARE 2018 statement: updating Consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 60 (2018) 132–136.
[5] A.S. Pappo, D.N. Shapiro, W.M. Crist, et al., Biology and therapy of pediatric rhabdomyosarcoma, J. Clin. Oncol. 13 (1995) 2123–2139.
[6] John R. Goldblum, Rosai and Ackerman's surgical pathology, Soft Tissues 41 (2020) 1810–1914.
[7] P. Dias, D.M. Parham, D.N. Shapiro, et al., Myogenic regulatory protein (MyoD1) expression in childhood solid tumors: diagnostic utility in rhabdomyosarcoma, Am. J. Pathol. (1990).
[8] W.M. Molenaar, J.W. Oosterhuis, A.M. Oosterhuis, F.C. Ramaekers, Mesenchymal and muscle-specific intermediate filaments (vimentin and desmin) in relation to differentiation in childhood rhabdomyosarcomas, Hum. Pathol. 16 (1985) 838–843.
[9] I. Van den Berghe, C. De Clercq, J. Casselman, Radiation-induced mandibular adult spindle cell rhabdomyosarcoma Flux RSS, Int. J. Oral Maxillofac. Surg. 37 (4) (2008) 395–397.
[10] S.M. Monte, G.M. Hutchins, G.W. Moore, Metastatic behavior of rhabdomyosarcoma, Pathol. Res. Pract. 181 (1986) 148–152, http://dx.doi.org/10.1016/S0344-0338(86)80003-6.
[11] Andrea Ferrari, Palma Dileo, Michela Casanova, Rossella Bertulli, Cristina Meazza, Lorenza Gandola, et al., Rhabdomyosarcoma in adults a retrospective analysis of 171 patients treated at a single institution, Cancer 98 (3) (2003) 571–580.