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Rhabdosarcoma of the posterior wall of the oropharynx: A case report

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Abstract
Sarcomas are rare malignant tumors with a mesenchymal origin. The sarcomas collectively represent approximately 1% of all adult malignancies and 15% of pediatric malignancies. It develops either from soft-tissues, including fat, muscles, nerves, vessels and conjunctive tissues, or from bones and leading to osteosarcomas. Head and neck soft-tissue sarcomas (HNSTS) represent approximatively 9 to 10 % of all soft-tissue sarcomas, and between the HNSTS, the oropharyngeal location is very rare. We present the case of a 47 years woman, operated for a T2N0M0 rhabdomyosarcoma of the posterior wall of oropharynx. The mass was burgeoning and obstructing the oropharynx of the patient. The surgical approach was trans-oral, the surgery tried to assure 1 cm security margins around the tumor attach and the histology found safe margins laterally and in depth. The patient had no postoperative radiotherapy after a multidisciplinary session's decision. The follow-up was of two years, clinically and radiologically, with no particular event, showing no local recurrence.

Keywords: rhabdomyosarcoma, sarcoma, oropharynx, woman, mesenchymal

Introduction
Head and neck soft-tissue sarcomas (HNSTS) represent approximatively 9 to 10 % of all soft-tissue sarcomas [1], knowing that all sarcomas represent approximately 1% of all adult malignancies [2]. The oncologic outcome of HNSTS is known to be worse than the outcomes of sarcomas from other body regions, because some histological forms of HNSTS show locally invasive and highly metastatic features [3]. Guidelines for management of sarcomas, even for HNSTS, exist, thanks to scientific societies work, like AJCC in their 8th edition of "AJCC TNM staging system for head and neck soft‐tissue sarcoma in adult patients" [3], or NCCN (national comprehensive cancer network) [4] or FNCLCC (Fédération Nationale des Centres de Lutte Contre Le Cancer) with their histological grading system proposed by the for tumor stratification [5]. According to HeeJung Kim in their study on HNSTS, oropharyngeal location represented 2 % of all HNSTS locations, and in this particular location, 26% were rhabdomyosarcomas [3].

Case report
We present the case of a 47-year-old woman, with no particular pathological history, seen in consultation for dysphagia in a 6 months progressive installation, and for the discovery by herself, of a mass behind her tongue base. The clinical examination found a burgeoning mass of the posterior wall of the oropharynx, painless, pedicle, partially obstructing the pharyngeal pathway, and no palpable cervical lymph nodes. A cervical CT-scan showed the presence of an expansive process of the oropharynx, measuring 23 x 21 mm, in hypo signal in T1 and T2, enhancing homogeneously after injection of contrast medium, and no cervical lymphadenopathy. The mass was therefore classified T2N0M0.
Biopsy of the tumor was performed, revealing after immunohistological examination a rhabdomyosarcoma. Trans-oral approach had been decided, and we proceeded to the tumor resection within 1-centimeter margins, arriving in depth to the prevertebral fascia. Despite the fact that the surgical field was narrow considering the approach, we managed to obtain 1-centimeter margins. The final histology showed a rhabdomyosarcoma, of low grade, according to the histological grading system proposed by the FNCLCC (Fédération Nationale des Centres de Lutte Contre Le Cancer) for tumor stratification. The surgical margins were safe; the decision of not proceeding to radiotherapy was decided in a multidisciplinary session.
The follow up was of two years till now. Our patient was seen every 3 month the first year, every 6 months the second year. We proceeded to MRI one year after surgery, where no suspect lesion was shown. CT-scan of lungs after 2 years was normal.

Discussion
Sarcoma is a rare malignant tumor of mesenchymal origin, consisting of heterogeneous groups of histology and sites of origin [2,3,5,6] More than 50 different histologic subtypes of sarcomas have been identified until now.
They collectively account for approximately 1% of all adult malignancies and 15% of pediatric malignancies. [1] We divide them classically into two groups: sarcomas of soft-tissues, which develop from fat, muscles, nerves, vessels or conjunctive tissue; and osteosarcomas. The most common primary sites are extremities (43%), the trunk (10%), viscera (19%), retroperitoneum (15%) [2]. Head and neck soft-tissue sarcomas (HNSTS) represent approximatively 10% of all soft-tissue sarcomas [3]. The location of the oropharynx between all HNSTS is rarer. According to HeeJung Kim and al. in their study on head and neck soft-tissue sarcoma (STS), oropharyngeal location represented 2 % of all HNSTS locations, and in this particular location, 26% were rhabdomyosarcomas. [3]
Adequate and high-quality imaging studies are important for clinical management of patients, and for preparing the

possible surgery. The quality of the imaging, and it good study, permit to have details about tumor size, about its contiguity to nearby visceral structures and it proximity to nerves and vessels. MRI is recommended, coupled or not with CT-scan. The MRI was in our case, the radiological exam we chose to study the mass, it showed us the relation of the tumor with posterior pre vertebral fascia, and local structures.

According to the 8th edition of clinical practice guideline by national comprehensive cancer network (NCCN), the first line of treatment recommended is surgical wide resection, and surgeons should secure safe resection margins as much as the anatomy of the area permits it. [4]. The margins should be 1 or 2cm, the anatomical barriers and adjacent structures have to be respected and the functionality should be well considered [4,7] We joined this surgical attitude by trying to assure 1cm margins despite the narrow space of surgery we had.

The same publication indicate that reconstructive surgery should accompany the surgical resection at the same time if possible, done by experienced head and neck surgeons in both oncologic and reconstructive surgery. For our patient, regarding to small attach of the pediculate tumor, we sutured the margins in a vertical way, with no tension and with a satisfying esthetic result [4]. (Figure 2)

Radiation therapy (RT) is used in case of positive resection margins and tends to improve the global prognosis of HNSTS [8]. In our case, regarding to our safe margins after final histological exam, and our tumor being classified as a T2N0M0, we decided after multidisciplinary staff, not to proceed to radiotherapy. This decision is in accordance with the guidelines of NCCN (national comprehensive cancer network), and the most-recommended histological grading system proposed by the FNCLCC (Fédération Nationale des Centres de Lutte Contre Le Cancer) for tumor stratification [4, 5].

Although it is uncommon among head and neck neoplasms, the oncologic outcomes of HNSTS is known to be worse than the outcomes for sarcomas from other body regions, because some histological forms of HNSTS show locally invasive and highly metastatic features [3]. In several studies, histologic grade and surgical resection margins were the most frequently suggested prognostic factors [9].

The final examination of our tumor showed a low-grade rhabdosarcoma, and the safe margins obtained laterally and in depth, make of this sarcoma a better prognosis

For the NCCN in their last guidelines publication, for a tumor classified stage IB, meaning the T2-T4, N0, M0, which is the case of our patient's sarcoma, rehabilitation should be consider until maximal function is achieved, the historical and physical examination should be done every 3–6 month for 2-3 years, then annually [4]. Concerning radiological follow-up, chest imaging and postoperative periodic imaging of primary site should be done, to be able to estimate the risk of locoregional recurrences [4]. The total follow-up period was 2 years: our patient was seen every 3 month the first year, every 6 months the second year. We proceeded to MRI one year after surgery, where no suspect lesion was shown. CT-scan of lungs after 2 years was normal. Angiosarcoma and rhabdomyosarcoma are known to recur more than did other subtypes, [10] that's reason why we are planning to be careful in the follow up of our patient.

Fig 1: Cervical CT-scan in sagittal section showing the rhabdomyosarcoma attached to the posterior wall of the oropharynx.

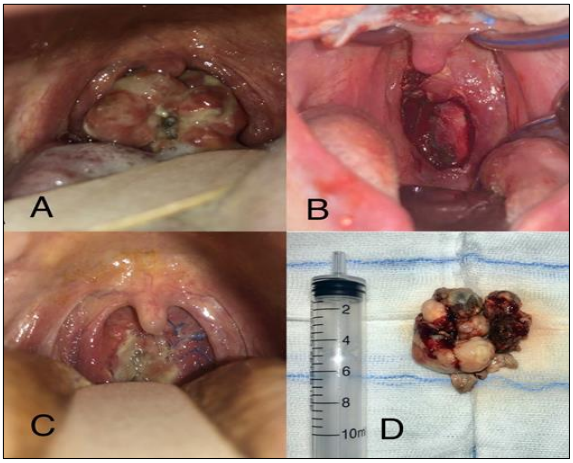

Fig 2: Macroscopic aspect of the rhabdomyosarcoma. A: burgeoning mass obstructing the oropharynx with an irregular aspect. B: per-operative view of the oropharynx after tumor excision. C: view of the oropharynx 6 days after surgery. D: resected tumor

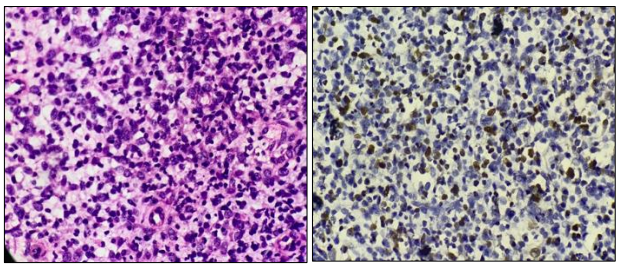

Fig 3: A: histology of the Rhabdomyosarcoma, hemateine eosine staining (microscopic ampliation x40) B: immunohistochemistry of the Rhabdomyosarcoma, nuclear marking (microscopic ampliation x40).

Conclusion

The rhabdosarcoma of oropharynx is a rare case, and fortunately for the patient, the diagnosis was not late, what permitted to have surgery with no heavy other treatment, and no complications. The early discover of this sarcoma permitted also to have a good prognostic for the follow-up of the woman.

Consent of the patient: The patient states that she does not have problems with the publication of her case

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