UNUSUAL PRESENTATION OF THE PLEOMORPHIC SARCOMA

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Abstract
The head and neck region is an uncommon site for pleomorphic sarcoma (PS). In the head and neck, PS has been reported mostly in nasal cavity and paranasal sinus accounting for 30% of all cases. The objective of this study is to report an unusual pleomorphic sarcoma in soft tissue of the neck.

Key words:
Pleomorphic Sarcoma, unusual presentation, head and neck.

Introduction
Pleomorphic sarcoma (PS), previously described as malignant fibrous histiocytoma, is the most frequent sarcoma of soft tissue in the late adulthood. The mandibulomaxillary region is the most common site for PS in the skull. The head and neck region is an uncommon site for pleomorphic sarcoma.

The terms were first introduced in the early 1960s to refer to a group of soft tissue tumors, characterized by a storiform or cartwheel-like growth pattern. The etiology of this tumor is unknown but seems to be multifactorial. Genetic background, environmental factors such as trauma, radiotherapy and malignant transformation from benign lesions can be involved in the etiology of this malignant tumor.

In the head and neck, PS has been reported to involve the nasal cavity and paranasal sinuses most frequently. Other reported sites involving the head and neck include the craniofacial bones, larynx, major salivary gland, oral cavity, pharynx, ear, eyelid, and cervical region. This report describes an unusual pleomorphic sarcoma in soft tissue of the neck.

Results and Discussion
REPORT: A 31 year-old female was referred to the head and neck surgeon for evaluation of a nodule in the neck with one year of development. She presented odynophagia for one week and night fever for one month. On extraoral examination, a swelling of the anterior region in the neck with multinodular surface, fibroelastic consistency, and mobile palpation as well as hyperemia in the center of the mass (suggesting a fistula) was observed. There was no evidence of cervical lymphadenopathy. Skin, mouth and oropharynx were without lesions. The patient was submitted to incisional biopsy and the initial diagnosis was PS. A computed tomography with intravenous contrast in an axial section, revealed a multinodular image with soft tissue density composed by confluent nodules. The image was located in subcutaneous of the left antero-lateral cervical region at level 3. The cricoid and thyroid cartilage (first and second tracheal ring) involving the coating of deep cervical fascia without invasion of adjacent tissue (Figure 1). A resection of the tumor was performed with oncological surgical margin and primary closure (Figure 2). The histopathological analysis showed a nodular mass which mainly consisted of spindle or stellate cells and atypical multinucleated giant cells. The cells were loose in myxoid stroma or eventually arranged in a storiform pattern. No other specific features of differentiation were observed (Figure 3). The patient was submitted to complementary radiotherapy and chemotherapy. After 12 months of follow up, the patient was free of disease.

Conclusions
The present report illustrates a rare, aggressive and complicated case of PS in the soft tissue from the neck. Although, PS is one of the most common sarcomas noticed in some studies with a large case of series showing a certain prevalence in this region. It may grow fast to big masses and can be associated with soft tissue invasion. A proper and quick diagnosis is fundamental for the prognosis.

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Conflict of interest disclosure
The authors declare that they have no conflict of interest.