Angiosarcoma pouco diferenciado primário de glândula salivar parótida: relato de caso

Primary poorly differentiated angiosarcoma of the parotid gland

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RESUMO
As neoplasias mesenquimais primárias das glândulas salivares são raras. Os autores descrevem um caso de angiossarcoma primário da glândula parótida em um homem com 84 anos.

UNITERMOS: Hemangiossarcoma, Glândula Parótida, Patologia, Antígeno CD34, Sarcoma, Imunoistoquímica.

ABSTRACT
Primary mesenchymal neoplasms of the salivary glands are rare. The authors report a case of primary angiosarcoma of the parotid gland in a 84-year-old male.

KEYWORDS: Hemangiosarcoma, Parotid Gland, Pathology, Cd34 Antigen, Sarcoma, Immunohistochemistry.

INTRODUCTION
Salivary gland tumors are uncommon tumors that exhibit an annual incidence around 0.4 to 6.5 cases / 100,000 people (1). Salivary gland tumors account for between 2.0 to 6.5 percent of all neoplasias of the head and neck (1,2,3). The peak incidence of this group of lesions occurs in the sixth and seventh decades of life, and the average ages of patients with benign and malignant tumors are about 46 and 50 years, respectively (1). Males and females are equally affected in general (1). The parotid gland is the most frequently involved site (around 65% to 80% of cases), in special the superficial (lateral) lobe, whereas the minor salivary and submandibular salivary glands are affected less commonly (25% to 10%) (1,2).

Between 55% to 70% of all tumors are benign epithelial lesions arising in the major salivary glands, and pleomorphic adenoma, basal cell adenoma and Warthin tumor are the most common histological types (1,4). A greater proportion of malignant tumors occur in the minor salivary gland than in major salivary glands, and mucoepidermoid carcinoma is the most common histological type (1,4,5). Mesenchymal tumors arising in salivary glands are rare lesions, with an incidence estimated in 1.9% to 4.7% of all tumors of these glands (1,6). Over 85% of mesenchymal tumors arise in the major salivary gland, especially in the parotid gland, and 75% to 80% of these cases are haemangiomas, with the greatest incidence occurring in the first decade of life (1,2,6). Salivary gland sarcomas are very rare aggressive tumors (0.3% of all salivary gland neoplasms) that arise in an older population than their benign soft tissue counterparts, with haemangiopericytoma and malignant schwannoma being the most frequent lesions (1,2,3,4,6).

Herein, the authors report a case of angiosarcoma arising in the right parotid gland, presenting the morphologic

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and immunohistochemical findings and diagnostic criteria of these this rare tumor.

**CASE REPORT**

A male patient, 84 years, caucasian, came to the outpatient hospital services referring as main clinical complaint the presence of a painless nodule in the topography of the right angle of the mandible, with slow growth for about a year. Physical examination revealed a firm nodule in the topography of the right parotid gland. Cervical lymphadenopathy or other head and neck abnormalities were not identified. The patient didn’t have prior history or clinical evidence of malignancy. CT scan of the head and neck revealed the presence of an intraparenchymal, circumscribed, ovoid tumor in the right parotid gland, which measured approximately 3.0 cm in greatest diameter. The initial procedure corresponded to total excision of the gland. The surgical specimen consisted of the parotid gland with perivisceral tissue, which weighed 9.0 g and measured 4.1 x 3.2 x 2.9 cm. On cute surface, it was found a reddish-brown, poorly delimited, firm, ovoid nodule, which measured 2.5 cm in greatest diameter. At microscopy, in parafin-embedded tissue, the lesion corresponded to a poorly differentiated highly cellular malignant neoplasm consisting of numerous small round cells, arranged in a solid or insular pattern, with high mitotic index and areas of necrosis (figure 1). The tumor did not infiltrate the capsule of the salivary gland or the perivisceral tissues. The immunohistochemical evaluation of the lesion showed positive expression for the antibodies CD34 (figure 2), CD31 and vimentin, and negative immunostaining for the antibodies AE1/AE3, CK7, CK20, CK8/18, CK14, EMA, CDX2, PSA, S100, TTF-1, desmin, actin, CD45 and CD10. The expression of the antibody Ki-67 was found in about 20% of cancer cells. The set of morphological and immunohistochemical findings corresponded, therefore, as a primary angiosarcoma of the parotid gland. The patient was referred for radiotherapy, and after four months of follow-up, showed no clinical evidence of local recurrence or systemic involvement.

**DISCUSSION**

Primary nonlymphoid mesenchymal neoplasms of salivary glands are uncommon tumors that account for only 1.9 to 4.7 percent of all neoplasms of this anatomic site (1,2,7,8). The majority of these tumors are benign, and hemangioma, lipoma, lymphangioma, and inflammatory myofibroblastic tumor are the most common histological types (1,2,7,8). Sarcomas arising in the major salivary glands represent about 0.3 percent of all benign and malignant tumors in this anatomic location, and about 1.5 percent of the malignant tumors (1,25,6,8). The criteria for establishing as a primary sarcoma arising in a salivary gland includes: 1) the patient must not have a similar sarcoma in other site; 2) the clinical evolution excludes the hypothesis of metastatic disease; 3) the gross and microscopic appearance of the lesion support a primary origin and excludes an invasive process arising in soft tissue; and 4) the diagnosis of carcinosarcoma have been excluded (1,2). Malignant peripheral nerve sheath tumors, rhabdomyosarcoma, and Kaposi sarcoma are the most frequent histological types (1,2,9).

Angiosarcoma arising in the parotid gland is a very rare malignant tumor, with a few cases reported in the literature (1,2,6,7). The lesion more commonly affects patients in the seventh decade (1,2,6,7). In general, the tumors are solid hemorrhagic nodular lesions, with irregular borders, measuring a few centimeters in diameter (1,2,6,7). At microscopy, the appearance of angiosarcoma is very variable, reflecting different degrees of vascular differentiation (1,2,3,7,9,10). Better differentiated tumors are characterized by irregularly infiltrative vascular channels, with a complex anastomosing growth pattern, lined by endothelial malignant cells showing various degrees of cytologic pleomorphism, nuclear atypia, mitotic index, and multilayering (1,2,3,7,8,9). In poorly differentiated tumors, there is a progressive loss of the vascular pattern, and the cells became more atypical, spindle-shaped, and close-packed, so that these cases can even resemble high-grade spindle cell neoplasias (1,2,3,7,8,9). Epithelioid angiosarcomas are made up of large rounded cells with relatively high nuclear grade that are arranged in small nests, sheets, cords or rudimentary vascular channels (1,11,12,13). Mitoses are easily identified, and hemorrhage and necrosis are common findings (1,2,3,7,8,9). Angiosarcomas usually express the usual vascular antigens including von Willebrand factor, CD31, and CD34 (1,4,11,12,13,14). Cytokeratin is expressed in about one third of soft tissue angiosarcomas (1,4,11,10,12,13,14).

![Figure 1 – Angiosarcoma of the parotid gland: a poorly differentiated neoplasia with small cells arranged in nests, HE, 200x. In detail, positive immunoexpression for CD34, streptavidin-biotin, 100x.](image)
Immunopositivity to actin and laminin can be found too (1,2,8). The differential diagnosis includes cases of poorly differentiated carcinomas, spindle-shaped and epithelioid sarcomas, metastases from malignant melanoma, and lymphomas (1,2,10,13,15,16).

The most successful treatments are wide surgical excision or surgery combined with radiation (1,2,6). The prognosis of primary angiosarcoma of parotid gland is uncertain (1,2,5,6,16). When located at soft tissue and skin, angiosarcomas are highly aggressive tumors (12,13). The features correlated with poor outcome include older age, invasion of adjacent structures, large size, and high Ki67 values (1,2,3,9,14,17). Histological appearance or grade does not correlate reliably with outcome (1,2,9,13,14). It looks that prognosis most strongly with clinical stage, emphasizing the importance of early diagnosis of small tumors (1,2,13). In this case report, the patient showed no evidence of recurrence after a follow-up of nine months, although a more aggressive clinical course was expected since it was a relatively large high-grade neoplasm with a high rate of cell proliferation.

FINAL COMMENTS

Herein, the authors describe a case of angiosarcoma arising in the right parotid gland of an 84-year Caucasian male patient. In general, angiosarcomas are uncommon soft tissue neoplasm with a predilection for skin and superficial soft tissues. In the absence of prior history of sarcoma, the diagnosis of primary angiosarcoma of parotid gland can be considered, and immunohistochemical studies are essential to establish the diagnosis in poorly differentiated lesions.

REFERENCES


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