

Primary Synovial Sarcoma of the Parotid Gland

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DOI: [10.36347/sjmcr.2020.v08i02.032](https://doi.org/10.36347/sjmcr.2020.v08i02.032)

| Received: 05.02.2020 | Accepted: 12.02.2020 | Published: 25.02.2020

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Abstract

Case Report

Synovial sarcoma is a rare malignant mesenchymal tumor of soft tissue. Its location in the parotid gland is exceptional. 55-year-old patient, with no specific history, with a history of slow evolution over one year of a mass in the right parotid region. The computed tomography objectified a tumor at the expense of the lobulated right parotid with serpiginous calcifications taking contrast on the periphery measuring 4 / 2.5 cm. The patient was initially treated by surgery and radiotherapy. The evolution was marked by good locoregional control without metastasis with a decline of four years. The diagnosis of parotid gland synovial sarcoma is confirmed by immunohistochemistry. Surgery combined to radiotherapy seems to be the best treatment.

Keywords: synovial sarcoma; parotid; surgery; radiotherapy.

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INTRODUCTION

Synoviosarcoma (SS) is a rare soft tissue mesenchymal tumor. It accounts for 7 to 10% of all soft tissue sarcomas; it affects the cervicofacial region in 0.7 to 9% (1). Its location in the parotid gland is exceptional. It affects most often the man, teenager or young adult (15 to 40 years) [1]. It is most often seen near the joints. Treatment is based on surgical excision followed possibly by radiotherapy or radio-chemotherapy (1). We report a case of synoviosarcoma of the parotid with a review of the literature.

CASE REPORT

Patient 55 years old, with no particular antecedents, with a history of slow evolution over one year of a mass of the right parotid region. The examination found a painless, mobile swelling measuring about 3 cm in its long axis of the right parotid region without facial paralysis. Computed tomography showed a tumor at the expense of the right parotid lobulated with serpiginous calcifications taking the peripheral contrast measuring 4 / 2.5 cm sitting on the outer portion of the gland at its superior pole (Figure 1).



Fig-1: CT of the parotid region in axial section after injection of the contrast medium: hypodense mass of 4cm / 2.5cm of the right parotid, lobulated with presence of serpiginous calcifications, taking the contrast at the periphery

An exo-facial parotidectomy with extemporaneous histological examination was performed, the result of which was suspect of malignancy. The intervention was then supplemented by a conservative total parotidectomy of the facial nerve with lymph node dissection of zones II and III. The final anatomico-pathological study microscopically revealed an invasive malignant tumor proliferation, comprising two quotas, the former made of short, monomorphous fusiform cells with an ovum, plump, hyperchromatic nucleus and a small cytoplasm; the

second was made of epithelial cells, the vascularization was of the hemangiopericytate type. In immunohistochemistry the epithelial cells expressed EMA with no expression of CKAE1 / AE3, AML and p63, the S100 protein was diffuse expressed. What concluded to a biphasic synovialosarcoma (figure 2, 3, 4), the examination of the ganglionic chains was N0.

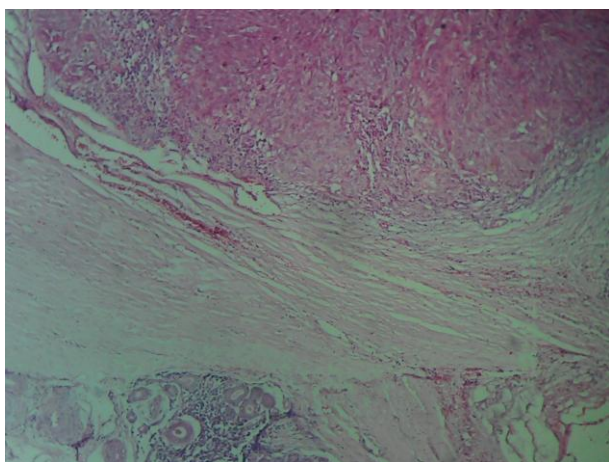


Fig-2: Tumor proliferation (top) of the parotid (bottom) (HEEx10)

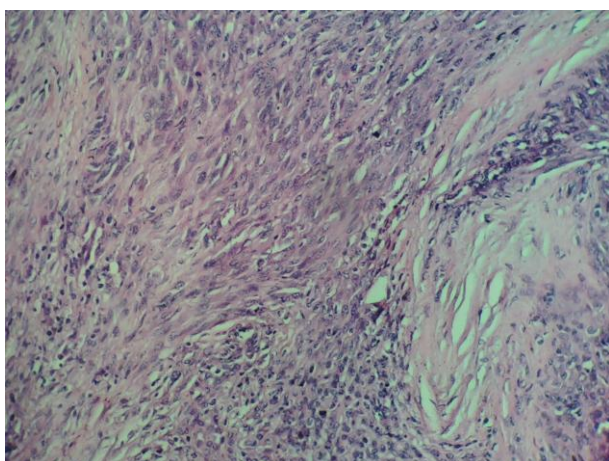


Fig-3: Fusocellular proliferation with hemangiopericytary like aspects (HEEx40)

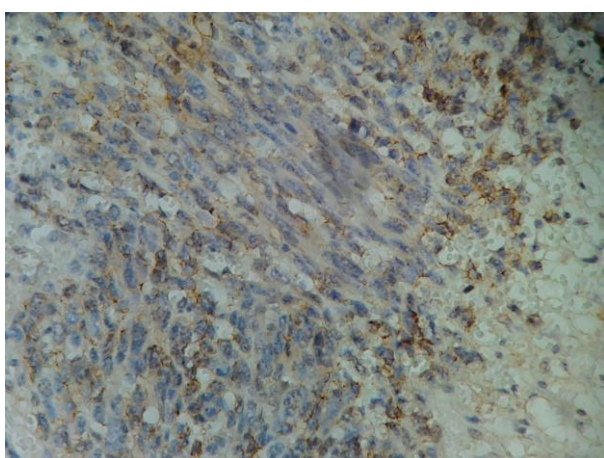


Fig-4: Moderate and focal positivity of the EMA (x100)

The patient had then received radiation therapy at a dose of 70 Gy spread over seven weeks based on 2Gy per day. The evolution was marked by a good locoregional control without metastases with a decline of 5 years.

DISCUSSION

Synovialosarcoma can occur at any age but most often affects young adults and adolescents with an average age of 38 years [2]. These tumors do not originate from synovial tissue, but rather from pluripotent mesenchymal cells near or even distant from articular surfaces [2, 3]. Synovialosarcoma sit more frequently in the extremities, particularly the lower limbs: thighs, knees, feet. The topography is often juxta-articular. In the head and neck region, the hypopharynx is the site most often involved because of the abundance of synovial tissue. The other cervicofacial sites are the parapharyngeal space, the naso-sinus area, and the pharynx [4]. The parotid localization is exceptional.

Without specific clinical signs, cervico-facial SS evolve slowly over several months [2]. Classically, patients consult for an indolent cervical mass gradually increasing in volume [1].

X-ray examinations are mainly of interest in tumor extension CT is more interesting for specifying bone erosion and intratumoral calcification, as for our patient, while MRI best specifies tumor extension, vascular invasion and intratumoral hemorrhage [3]. The diagnosis of certainty is histological, macroscopically the SS is presented as a rounded, well-defined, pseudo-encapsulated, and lobulated or plurinodular mass, sometimes with cystic and hemorrhagic foci at the sectional slice. Microscopically, two forms of SS are distinguished: a classical biphasic form and a rarer monophasic form [4, 5].

The biphasic form is characterized by the coexistence in variable proportion of two components, one epithelial glanduliform and the other fusocellular fibrosarcomatous appearance. In immunohistochemistry, epithelial cells express epithelial markers (cytokeratin (KL1), EMA); fusiform cells express the anti-vimentin antibody. The CD 99 and the PS 100 mark the two contingents in 50% of the cases in a focal and not extensive way and sometimes the epithelial contingent can express the ACE and (Human milk fetal globule protein) [2]; bcl2 is positive in 80% of cases. Some authors report an aberrant positivity to caldesmone and calponin. Whatever their type, most SS have translocation t (x18) (p11.2; q11.2) [4-6].

The SS can pose problems of differential histological diagnosis with other tumors of which the main ones are: carcinosarcoma, metastatic adenocarcinoma, fibrosarcoma, malignant schwannoma, hemangiopericytoma, and mixed salivary tumors (pleomorphic adenoma or myoepithelium) [4, 5].

Given the limited number of cases of parotid synovial sarcomas in particular and cervico-facial locations in general, there is no therapeutic standard. Cases reported in the literature have been treated by surgery with or without radiotherapy and ifosfamide-based chemotherapy in some cases.

Surgical excision remains the basic treatment. This excision should be wide with a margin of two cm macroscopically healthy tissue in all plans, to reduce the risk of local recurrence, sometimes at the expense of functional prognosis [4].

What is not always easy in the cervicofacial area, considering the anatomical considerations, The obviously ganglionic, in matter of cervico-facial SS, is recommended only in case of palpable lymphadenopathy, seen the character very little lymphophile of these tumors [1].

Radiotherapy is used postoperatively to improve local control, especially if the excision margins were not wide enough, the recommended dose is 65 Gy or more [4].

Chemotherapy with high doses of ifosfamide used as an adjuvant or neo-adjuvant may increase the rate of local control, but without impact on survival [11].

The prognosis of these tumors is reserved because of the frequency of metastases, most often pulmonary: 37% of cases and local recurrences, survival is about 50% at 5 years. Prognostic factors are size, tumor differentiation, extent of necrosis, mitotic index, tumor-free surgical excision margins [2, 8]. However, the prognosis has been improved by current therapeutic protocols including adjuvant radiotherapy even in the case of healthy margins.

CONCLUSION

The primary synovial sarcoma of the gland is extremely rare. His diagnosis is mainly histological.

Expanded surgery with margins remains the treatment of choice.

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