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CASE REPORT

Synovial sarcoma of the parotid gland – case report

Mięsak maziówkowy ślinianki przyusznej – opis przypadku

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ABSTRACT: Neoplasms of the salivary glands represent about 3% of the head and neck tumors, with the parotid gland being most commonly affected (about 80% of all cases). Pleomorphic adenoma is the most common tumor type, accounting for 80–90% of all diagnoses. On the other side synovial sarcoma is rare in the head and neck region, accounting for about 3% of all diagnoses, and it mainly occurs in the lower extremities. To date, about 20 cases have been described in this region. The etiology of the disease remains unknown, but there are reports linking synovial sarcoma with rearrangements in the gene responsible for chromosomal transcription t (X; 18) (p11, q11).

KEYWORDS: salivary glands, synovial sarcoma, parapharyngeal space

STRESZCZENIE: Nowotwory gruczołów ślinowych stanowią około 3% wszystkich nowotworów okolicy głowy i szyi. Zmiany te zwykle dotykają ślinianki przyusznej (ok. 80% wszystkich przypadków). Najczęstszym z nowotworów jest gruczolak wielopostaciowy (80–90%). Wśród rzadkich zmian nowotworowych znajduje się mięsak maziówkowy, który zwykle zlokalizowany jest w okolicy stawów kończyn dolnych. W przypadku głowy i szyi stanowi on zaledwie 3% wszystkich rozpoznań. Dotychczas opisano około 20 przypadków mięsaka maziówkowego zlokalizowanego w śliniance przyusznej. Etiologia choroby pozostaje nieznana, istnieją jedynie doniesienia wiążące mięsaka maziówkowego z rearanżacją w obrębie genu kodującego transkrypcję chromosomalną t (X; 18) (p11, q11).

SŁOWA KLUCZOWE: gruczoły ślinowe, mięsak maziówkowy, przestrzeń przygardłowa

INTRODUCTION:

Neoplasms of the salivary glands represent about 3% of all head and neck tumors. The parotid gland is the mostly affected salivary gland (about 80% of all cases), followed by intraoral minor salivary glands and the submandibular gland [1]. The most common neoplastic lesion is pleomorphic adenoma (tumor mixtus), accounting for about 80 to 90% of all cases. Malignant lesions are rarely found and account for about 5% of salivary gland neoplasms, of which synovial sarcoma (SS) is among the rarest types. It mostly presents in the lower extremities [2]; however, it also accounts for 3% of all head and neck malignancies. It mostly affects the parotid salivary gland, with about 20 cases described in the literature to date. We report a case of a 37-year-old male who presented with a synovial sarcoma in the left deep lobe of the parotid salivary gland.

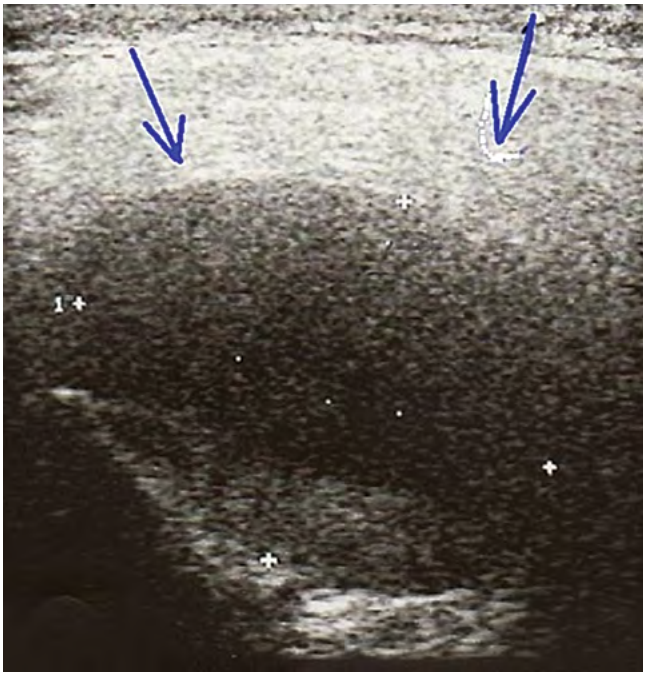

Fig. 1. Ultrasound examination revealing the tumor (arrows).

CASE REPORT:

A 37-year-old patient presented with an enlargement of the left parotid salivary gland and was admitted to our department. The enlargement appeared six months before admission, and it was not painful and slowly growing.

In order to evaluate the enlarged parotid salivary gland, an ultrasound scan was conducted. It revealed a well-delimited heterogeneous focal lesion, sized 29×24mm, in the central part of the pulp of left parotid gland, near the surface of the bone. The lesion was scarcely vascularized. The sonographic picture suggested an inflammatory, enlarged lymph node or a different characteristic change. The left parotid salivary gland was not enlarged, but it was modeled by the focal lesion. Local lymph nodes were not enlarged (Fig. 1).

Fine needle aspiration biopsy of the tumor under ultrasound guidance was performed, and it revealed protein deposits, dispersed epithelial cells, and groups of cells with granulated cytoplasm. Atypical cells were not present. A suspicion of mixed neoplasm of the parotid salivary gland was made.

Before admission, computed tomography was conducted. Medially, there was a hypodense (40jH), well-restricted mass, sized 38×25×23mm, extending from the temporomandibular joint; it had peripheral contrast enhancement; anterior subluxation of the left temporomandibular joint was visible. No infiltration in the surrounding soft tissues and bones was seen. The mass pushed the retromandibular vein posteriorly (Fig. 2). Cervical lymph nodes were not enlarged. Pharyngeal and laryngeal structures as well as the soft tissues of the neck were normal. The right mandibular sinus and ethmoid cells on the right side were tortuous, and a small polypoid mucosal swelling was seen in the left maxillary sinus.

On magnetic resonance imaging (MRI), there was a solid, expansive, slightly heterogeneous, abnormal mass, sized 50×50×40 mm in the inferior pole of the left parotid salivary gland. The mass enhanced with contrast, was well-delineated but without a clear capsule, and penetrated to the left infratemporal fossa. Swollen lesions were visible in the subcutaneous tissue, without infiltration. On both sides of the neck, numerous lymph nodes were enlarged to the size of 18×10mm. They were more numerous on the left side. There was a significant parietal cystic mucosal swelling in the maxillary sinuses (Fig. 3).

On admission, the patient was in a good overall condition. The lesion in the parotid salivary gland was hard, solid, with restricted mobility, and not painful on palpation. Facial nerve function was preserved. Laboratory findings and chest x-ray were normal.

Apart from arterial hypertension, the patient's medical and surgical history was unremarkable. He was qualified for surgery. The tumor was removed from the parapharyngeal area under neuromonitoring of facial nerve function. The function of the facial nerve was preserved.

On histopathology, the tumor appeared as a malignant spindle-cell neoplasm, probably of mesenchymal origin. The tumor was hypercellular, with numerous mitoses and areas of necrosis. The cells were small and had features of high-grade atypia as well as focal pseudoepithelioid features. On immunohistochemistry, the cells were largely positive for vimentin, CD99, CD56, and bcl2; additionally, several cells were positive for broad spectrum cytokeratins. The neoplastic cells were negative for desmin, SMA, S100p, CD34, and CD117. The pathological report indicated a soft tissue malignancy with features of synovial sarcoma, and it recommended fluorescent in-situ hybridization analysis for t (X, 18) (p11, q11) (Fig. 4). Molecular verification confirmed a rearrangement in the SS18 gene and proved that the neoplasm of the parotid salivary gland was a synovial sarcoma. In 65% of the analyzed cells, there was one not separated and one separated signal. In 15%, there were two not separated signals. In 14%, there was one not separated signal and one signal 3' SS18 or one signal 5' SS18. 18. 6% of cells had two not separated signals and one separated signal.

Before further treatment, the patient was referred for PET--CT. It revealed an expansive mass in the region of the left temporomandibular joint. It was metabolically active, sized 35×12×26mm, with SUVmax = 4.9 mm. The lesion penetrated into the left parotid salivary gland. Increased FDG metabolism was found in the vocal folds (SUVmax=5.3 mm), and no visible lesions were found on CT, suggesting that the lesion was only functional. No other focal metabolic lesions in the head and neck were observed. No metastases were seen.

The patient was referred for adjuvant chemotherapy, and high doses of iphosphamide (3 × 750 mg) were recommended. However, the patient refused to receive the third dose.

At 5-month follow-up, contrast-enhanced MR examination was performed. It revealed local recurrence in the left parapharyngeal space, masseter space, parotid gland, and pterygopalatine fossa. Retromandibular lipoid tissue was filled with the tumor mass. Pterygoid muscles and masseter muscle were infiltrated by the tumor. The lesion size was 68×52×64mm, it was polycyclic, well-restricted, and showed contrast enhancement. The tumor infiltrated the base of the middle cranial fossa and meninges; it penetrated to the base of the middle cranial fossa and modeled the basal part of the temporal pole. No infiltration in the temporal lobe was noted, but small satellite lesions were observed. Under the main tumor mass, there was a solid mass (45×38mm) with a different signal and contrast enhancement pattern than the main lesion; possibly, it represented lymph nodes. Around the tumor, swelling of muscles and subcutaneous tissue was present.

The patient was referred for another surgery. Left parotidectomy with ligation of the external carotid artery was performed under general anesthesia. The tumor was removed with a segment of the external carotid artery, and meninges of the base of the middle cranial fossa. The external carotid artery was underpinned below the tumor. The tumor site after resection was filled with lipoid tissue that had been taken from the abdominal wall. Additionally, lymph nodes from the third field were removed. Surgery and hospitalization were uncomplicated. On discharge, facial nerve function was assessed at 4/6 H-B.

On histopathology, the mass had fragments of a salivary gland, connective and lipoid tissue, skeletal muscle with numerous focal infiltrative lesions of a spindle-cell neoplasm. The neoplasm had high mitotic activity (about 35 mitoses/10 FOV). Moreover, a small spread of necrotic lesions was seen. On immunohistochemistry, the lesion was BCL2-positive, CD99--positive, CD34-negative, S100-negative, and SMA-positive. Additionally, few cells were desmine-negative or CKAE1/E3--positive. Eleven lymph nodes presented reactive lesions. Taking

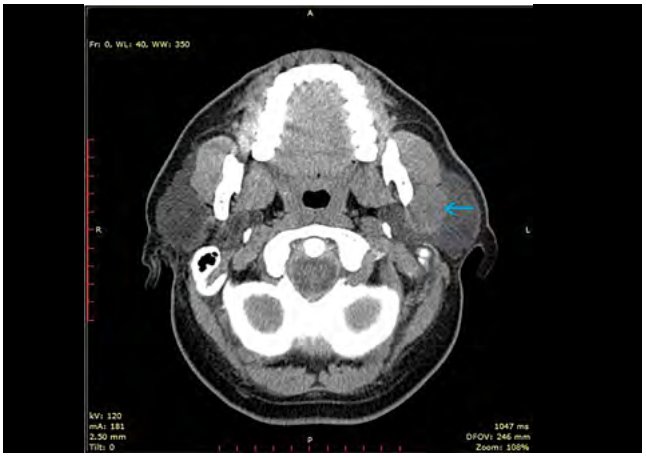

Fig. 2. CT shows the tumor (arrow).

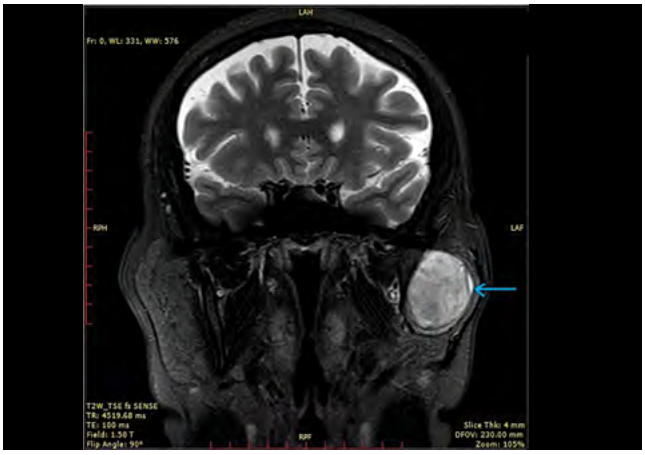

Fig. 3. MRI reveals the tumor (arrow).

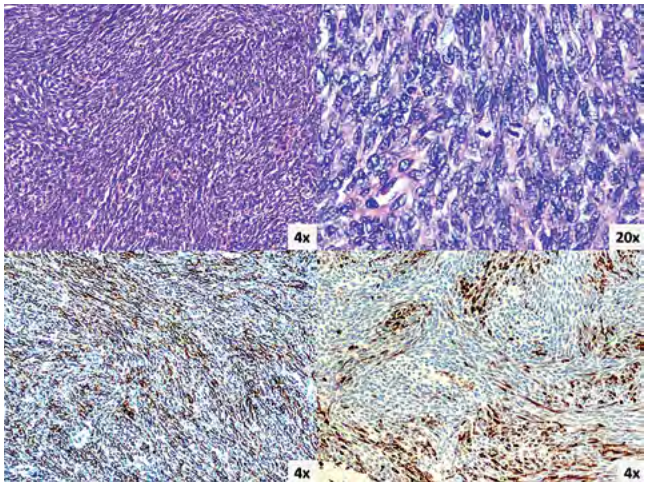

Fig. 4. Upper row - microscopic view of the spindle-cell and small-cell tumor with epithelioid features and numerous mitoses. Magnification of 40x and 200x, respectively Lower row - tumor cells are largely positive for CD99 and are focally positive for broad spectrum cytokeratin. Magnification of 40x.

into consideration the morphological picture and additional results, infiltration of synovial sarcoma should be considered along with fibrosarcoma.

Six weeks after surgery, PET-CT was performed. In the head and neck region, no focal lesions with increased accumulation of FDG were noticed. The post-surgical lodge on the base of the skull was filled with lipoid tissue. On the left side of the neck near the excised parotid salivary gland, dense tissues were observed. They presented little, spilled metabolic activity (SUV max up to 3.4mm). There was a small, band-like tissue density extending along the sternocleidomastoid muscle with a similar metabolic activity (SUVmax up to 3.3 mm). Possibly due to post-operative changes. No lymph nodes were metabolically active. No metabolic metastases were seen.

The patient was qualified for radiotherapy. The irradiated area included the temporomadibular region, the base of cranium, and left parotid region. The total dose was 66Gy (fractionated doses of 2Gy).

DISCUSSION:

Synovial sarcoma in the head and neck region is a rare disease, as only about 40 cases have been described to date. The mostly affected gland in this region is the parotid salivary gland.

In our case, the lesion was found in the deep lobe of the left parotid gland, penetrating to the parapharyngeal space. The most common neoplasm in the parapharyngeal space is pleomorphic adenoma. Synovial sarcoma (SS) is the fourth most common sarcoma type after malignant fibrous histiocytoma, liposarcoma, and rhabdomyosarcoma [3].

Synovial sarcoma is a rare and aggressive soft tissue tumor, and it accounts for 7–8% of all malignant sarcomas [4]. The head and neck region is uncommon for that neoplasm, as it mainly occurs in the lower extremities [5]. Two types of SS are described in the literature, i.e., monophasic (consisting only of spindle cells) and biphasic (consisting of spindle and epithelioid cells).

Parotid gland synovial sarcomas occur predominantly in men (male-to-female ratio of 2:1) before the fifth decade of life [6].

On genetic examination, synovial sarcoma is characterized by rearrangements in the SS18 gene that encodes chromosomal transcription factors. There is a translocation involving sex chromosomes and chromosome 18 [t(X;18)(p11.2;q11.2)] [7].

In the presented case, a slow growth of the neoplasm was observed. Due to the localization of the tumor, a visible tumor could be observed late in the course of the disease. The patient did not complain of any pain, and there was no facial nerve palsy. As described by Ruggiero, tumors are commonly painless masses that grow asymptomatically for weeks [8].

FNAC conducted before the operation did conclusively rendered the diagnosis. The diagnostic value of FNAC is still discussed. According to the available literature, the compatibility of FNAC results with those of postoperative histopathologic examination is estimated at 85% [9]. There are also reports that do not recommend performing FNAC due to its low diagnostic value and a possibility of tumor spread during biopsy [10]. In our department, CNB and FNAC are performed routinely in neoplasms of salivary glands.

Complete surgical excision is the treatment of choice for tumors in the parapharyngeal region. Also, in the case of sarcomas, the treatment is mainly surgical. Postoperative treatment is dependent on the histopathologic examination of the removed tumor. In the case of sarcoma, local radiotherapy is suggested. Although SS is thought to be a chemosensitive tumor, the role of adjuvant chemotherapy remains controversial [11]. In the described case, high-dose iphosphamide was prescribed.

A high risk of distant metastases is an indication for a thorough analysis including PET-CT.

Based on the current data, it is estimated that 10- and 15-year disease-free survival ranges from 45 to 50% [12].

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