

Triton tumor of the parotid area. Case report

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Summary. A 27-year-old woman with a Malignant Triton Tumor (MTT), or malignant schwannoma with rhabdomyoblastic differentiation, located in the parotid cell and infiltrating the nasal sinuses and the left orbit is described. The initially resected tumor showed three recurrences within a 2 years follow-up period. During successive recurrences an increase in cellular density, number of mitoses and necrosis was noticed. Immunohistochemical analysis showed that the tumor was composed of a mixed population of cells. Some of them showed positivity for actin, desmin and myoglobin, while others were positive for S-100 protein, glial fibrillary acid protein, and IV-collagen. Cytokeratin stainings were negative. Up to now, 8 benign triton tumors and another 45 cases of MTT have been described. None of them was primarily located in the parotid gland, and infiltration to the orbital cavity has not been previously described.

Key words: Malignant triton tumor, Rhabdomyosarcoma, Malignant schwannoma, Parotid, Orbit

Introduction

The term «Triton tumor» is applied to any neoplasm sharing both neural and skeletal muscle differentiation. Benign forms are considered to be neuromuscular hamartomas, composed of mature nerve fibres and well-differentiated striated muscle fibres (Lagace, 1987). Only eight of these cases have been reported in the literature (Markel and Enzinger, 1982). The malignant counterpart (MTT) are therefore malignant peripheral nerve sheaths tumors (MPNST) with a rhabdomyosarcoma component. It is well known that about 15% of the MPNST show metaplastic areas with tissues such as epidermoid epithelium, melanocytes or heterologous elements like bone or cartilage. However, when the presence of scattered well differentiated skeletal muscle cells is obvious, the term MTT is applied. Immunohistochemical analysis with S-100 or NSE characterize the neural component of these tumors, while actin, desmin

or myoglobin characterize the rhabdomyoblastic component.

To the best of our knowledge, 45 cases have been previously described. Interestingly, none of them was initially located in the parotid area and infiltration to the orbital cavity has not been previously described (Dewitt et al., 1986; Shajrawi et al., 1989; Wong et al., 1991). Therefore, we consider the present case as interesting to be reported.

Materials and methods

A 27-year old woman, allergic to the iodine products, without any other interesting antecedents nor any von Recklinghausen's disease stigmata, attended another hospital in 1992 because of the presence of a 3.5x3 cms spheroidal mass affecting the left parotid surface lobe.

A thin-needle aspiration puncture and a subsequent biopsy were performed. An initial «neuroectodermic malignant tumor» diagnosis was established at that point, whilst waiting for immunohistochemical results.

In March, 1993, she received two chemotherapy cycles and 10 Co-therapy up to an amount of 30 Gy (3000 rad). The tumor reappeared, and in October, 1993 was treated with 70 Gy (7000 rad) of Co-therapy. In the following months the neoplasia infiltrated the nasopharyngeal space and the maxillary sinus, causing the transference of the patient to our center in January, 1994. Then a radical parotidectomy was performed opening the sinus and following the Caldwell-Luc procedure.

Eight months later she suffered painful left exophthalmos. The computerized tomography scan showed an infiltrative mass occupying the parotid cell, paranasal sinuses and left orbit (Fig. 1). She was then subjected to radical surgery with orbital exenteration (Fig. 2).

The resected specimen was routinely processed and stained with H&E, phosphotungstic acid-hematoxylin, Masson's Trichrome stain and Wilder reticulin stain. Immunohistochemical stainings were performed for skeletal muscle actin (HHF-35), myoglobin, desmin, S-100 protein, neuron-specific enolase, glial fibrillary acidic protein, myelin-associated glycoprotein, IV-collagen and cytokeratins using conventional Avidin-Biotin immunohistochemical techniques to elucidate the



Fig. 1. CT-SCAN showing the tumoral infiltration into the left orbital cavity and adjacent anatomic regions.



Fig. 2. Postsurgical CT-SCAN, taken after the wide skull-facial surgical removal.

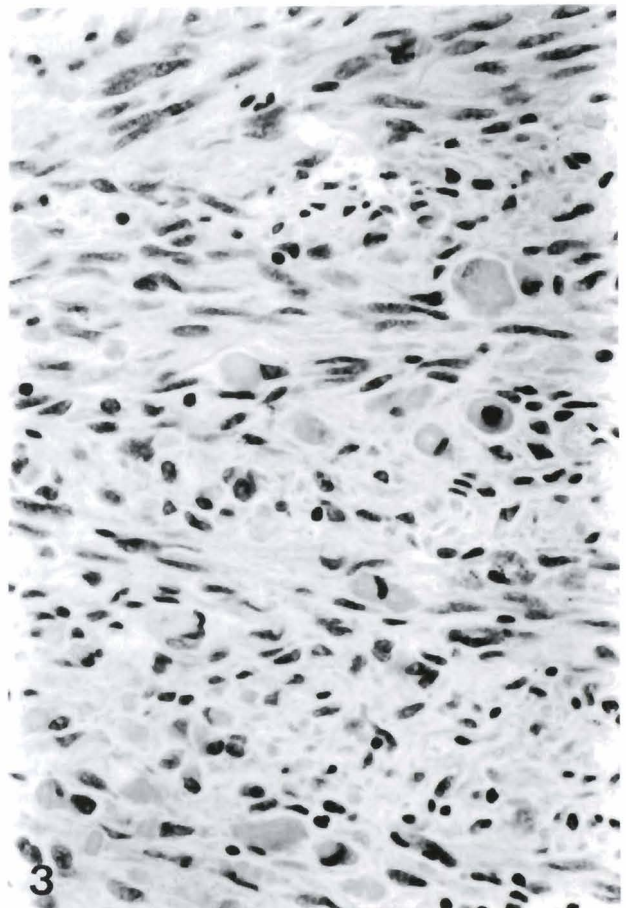


Fig. 3. Microscopic aspect of the tumor in the first specimen, where the tumor biphasic component can be observed. H&E. x 200

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mixed tumor cell population origin and to exclude other sarcomas and carcinomas that should show a different immunohistochemical pattern. To evaluate the mitotic index the number of mitosis in 10 high magnification areas was counted within representative tumor areas.

Results

Macroscopic examination of the 1st specimen revealed several irregular fragments of parotid gland and adjacent tissues. The 2nd specimen was a 10x8x7 cm mass including the left ocular globe and several maxillar bone fragments.

Microscopic examination of the tumor showed a double cell population: spindle cells with poorly defined cytoplasm mixed up with polygonal stretched cells, rich in eosinophilic cytoplasm (Fig. 3). In the latter cells, the presence of striated material was demonstrated with phosphotungstic acid-hematoxylin. Thus, some areas showed a nodular configuration (Fig. 4). The 1st specimen revealed a mitotic index of 1 mitosis per 10

hpf, while the 2nd one, in the highest cellularity areas, showed more than 5 mitoses per 10 hpf (Fig. 5). The histological sections corresponding to the following samples presented necrotic areas and interstitial hemorrhages.

In the sinonasal mucosae, neoplastic cells were more spheroidal, less differentiated, infiltrating the corion and forming slack polypoid masses covered by respiratory epithelium (Fig. 6). The neoplastic cells infiltrated bone structures, connective tissues of the orbital cavity and the episclera, not invading the ocular globe nor the optic nerve. Histological sections of these areas showed more and bigger necrotic areas than the primitive tumor. An epithelioid growth pattern was also noticed (Fig. 7).

Immunohistochemically, both specimens showed positivity for actin and desmin (Fig. 8), diffuse positivity for S-100 protein and glial fibrillary acid protein. Thus, isolated cells in a scattered pattern, showed positiveness for myoglobin and myelin-associated glycoprotein. IV-collagen was only shown in the 1st specimen. Cytokeratin staining was negative in both specimens.

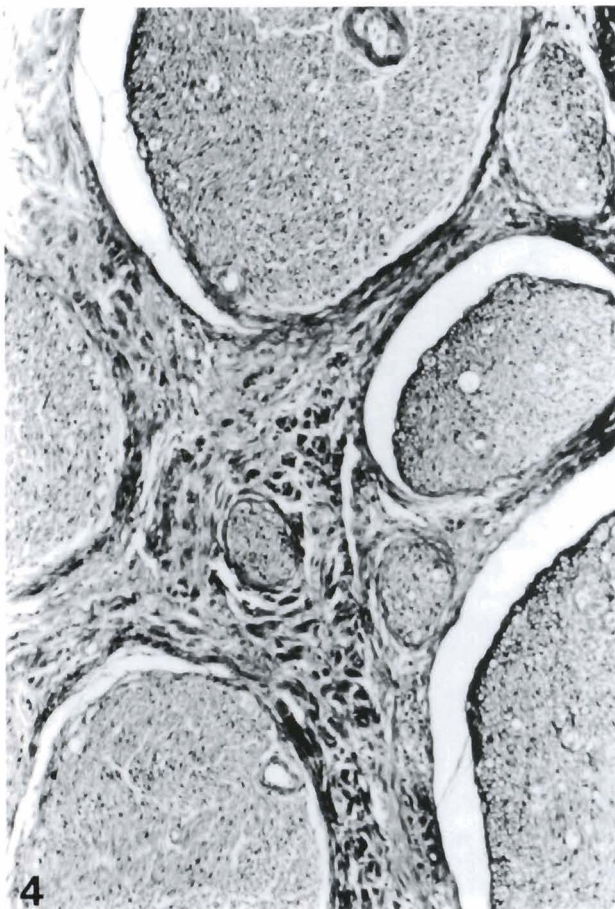


Fig. 4. Nodular pattern area with neuroid appearance. Masson's Trichrome connective tissue stain. x 125

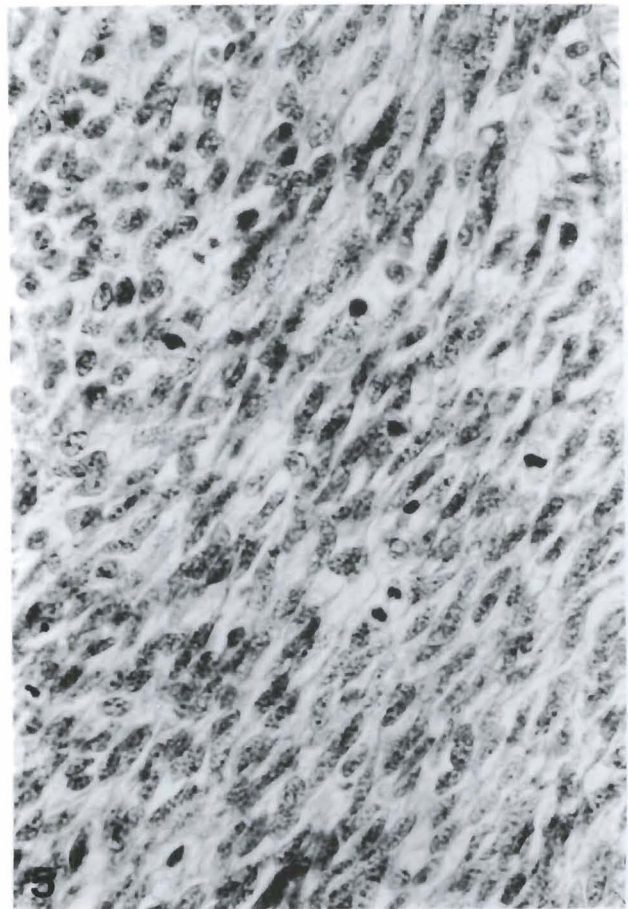


Fig. 5. The MTT in the 2nd specimen shows higher cellular density areas and a high mitoses number. H&E. x 250

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Discussion

In previous subject reviews performed by Brooks et al. (1985), Dewit et al. (1986), Bhatt et al. (1991) and Heefner and Gnepp (1992), the authors did not find any MTT affecting the parotid cell or the orbitary cavity. However, MTT involving the temporal fossa (Ducatman and Scheithaner, 1983), the paranasal sinus (Shajrawi et al., 1989), the palate (Shotton et al., 1988), the thyroid (Boss et al., 1991) and the acoustic nerve (Han et al., 1992) have been reported. Indeed, no previous MTT reference has been found affecting major salivary gland regions. Looking for related tumors involving salivary glands, Seifert et al. (1986) and Seifert (1992) reported 5 cases of malignant Schwannomas and 4 cases of embryonal rhabdomyosarcomas; Luna et al. (1991) reported 2 cases of both neurosarcomas and rhabdomyosarcomas; Auclair et al. (1986) 11 cases of malignant Schwannoma; and finally, Pescioli et al. (1986), a case of malignant Schwannoma of the submandibular gland.

The histogenesis of these unusual tumors has been controversial since they were first reported by Masson and Martin in 1932 (Woodruff et al., 1973; Woodruff,

1993; Woodruff and Perino, 1994). Originally, skeletal muscle differentiation was suggested to be induced by the neural elements, in a similar way was the normal nerve was believed to induce skeletal muscle regeneration in the Triton salamander. Recent studies have shown that stem cells migrating from the neural crest can show divergent differentiation towards mesenchymal elements so therefore, cells of tumors originating from those stem cells can have both the characteristic rhabdoid pattern or schwannoid spindle cell features (Woodruff, 1993). Immunohisto-chemical analysis revealed both the neural and muscular differentiation of the tumor cells as previously mentioned. Moreover, the coexpression of S-100, vimentin and desmin indicates a common neuroectodermic origin (Daimaru et al., 1984; Schmidt et al., 1990; Smirnov and Posvisil, 1990).

In this reported case of MTT some remarkable clinico-pathological features should be mentioned. An exhausting clinical examination of the patient did not reveal any von Recklinghausen's disease stigmata. Therefore, the present case should be considered as a sporadic case, corresponding to Group II in the Brooks et al. (1985) classification. As previously described,

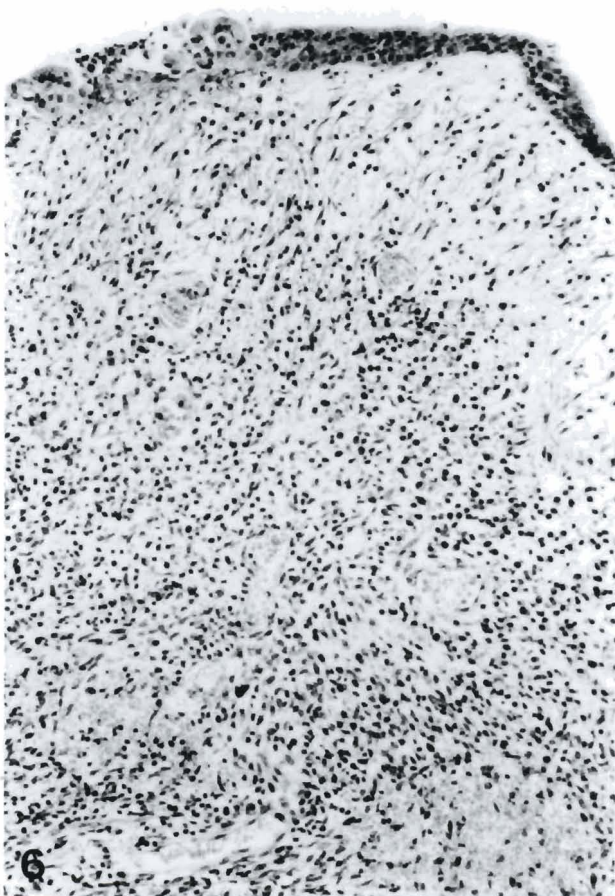


Fig. 6. Maxillar sinus mucosa infiltrated by MTT Producing polypoid masses. H&E. x 125

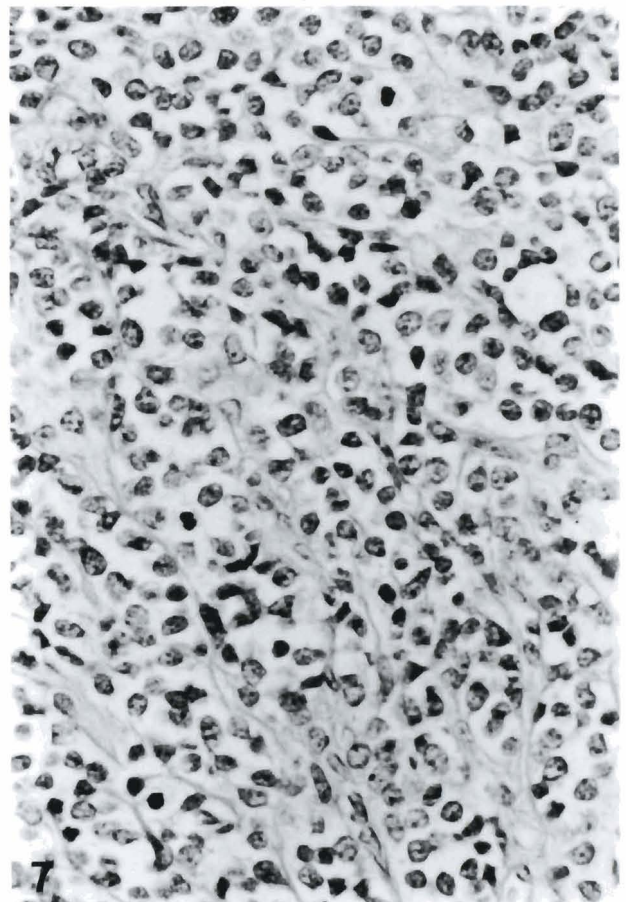


Fig. 7. MTT epithelioid pattern at orbitary level. Masson's trichrome connective tissue stain. x 250

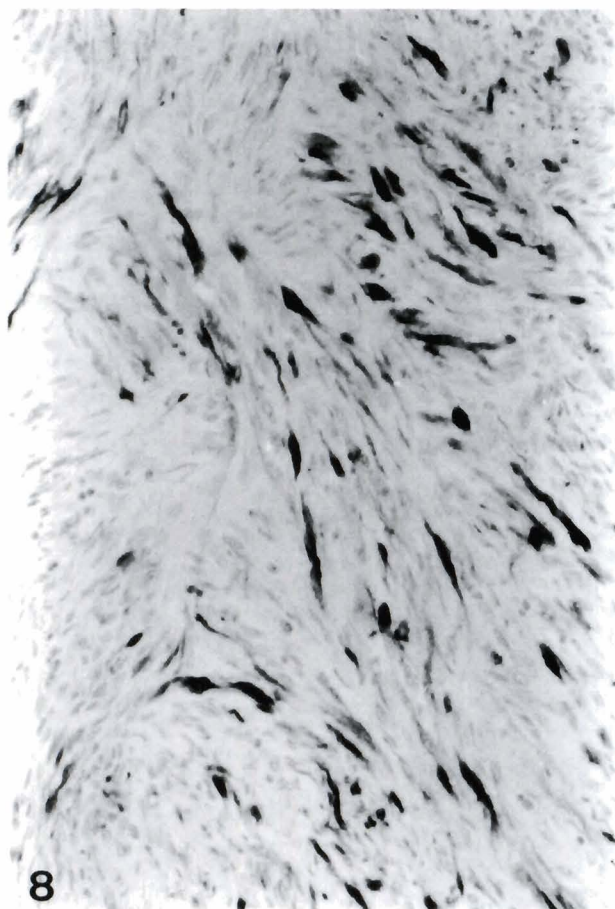


Fig. 8. Immunohistochemical study showing the coexistence of neural cells and cells with rhabdomyoblastic differentiation. Desmin. x 200

tumor cells adopted different architectural patterns in different tumor locations: parotideal, nasopharyngeal, sinonasal and orbital cavity. Thus, an unusual epithelioid pattern was noticed in the last recurrence. The epithelioid variant of malignant Schwannoma has been fully described by Laskin et al. (1991) and in sinonasal tumors by Fernández et al. (1993), occasionally with melanocytic differentiation (Karcioglu et al., 1977; Cardesa et al., 1985; Ooi et al., 1992).

The biological behaviour of MTT, with high local aggressiveness, high recurrence rates and a metastatic capability, makes a wide surgical resection with free surgical margins necessary at the moment when the initial diagnosis has been made (Awasthi et al., 1991). Chemo- and radiotherapy are not effective, and although postsurgical complementary radiotherapy is recommended, it is not reliable. Indeed, the reported patient presented a remarkable sialoadenitis after the irradiation, together with a severe loss of parotid parenchyma, while the tumor mass was mainly preserved with few necrotic areas. This patient's MTT increased its cellularity and number of mitosis in the last recurrence. These remarkable features should always be

considered by clinicians to improve the poor prognosis and low survival rates of these patients (Heffner et al., 1992).

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