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CASE REPORT

Massive infra-clinic invasion of the facial nerve by a myoepithelial carcinoma of the parotid

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Summary We report a new case of myoepithelial carcinoma of the parotid gland in an 8-year-old girl. This is the first case published in a child. The parotid tumour was slightly tender and measured almost 2 cm in diameter. There was no associated facial nerve paralysis despite surgical and histologic evidence of massive facial nerve infiltration. We performed total parotidectomy with resection of the intra-mastoid portion of the facial nerve completed with prophylactic lymph node dissection. Eight months after surgery, MRI revealed a deep-lying recurrence, which required reintervention. There has been no subsequent recurrence 18 months after surgery. Microscopic examination of operative specimens confirmed the diagnosis of parotid myoepithelial carcinoma with fusiform cells. Immunohistochemical markers were positive for cytokeratin, epithelial membrane antigen, smooth muscle actin, S-100 protein, anti-desmine and anti-vimentine. This difficult to diagnose tumour, which was individualised by the World Health Organisation in 1991, is considered a moderate to high-grade malignancy when it develops in a pleomorphic adenoma or appears de novo.
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1. Introduction

Myoepithelial carcinoma of the parotid gland is a rare tumour representing less than 0.5% of all salivary gland tumours. First described by Sheldon [1] in 1943 who reported three cases discovered in pleomorphic adenomas, it was not until 1991 before the World Health Organisation officially included this denomination in the histologic classification of salivary gland tumours. These invasive tumours are constituted by atypical myoepithelial cells with an increased mitotic index, thus differentiating them from benign myoepitheliomas [2]. Only 75 cases have been published and the majority involved single cases. Accordingly, its clinical and
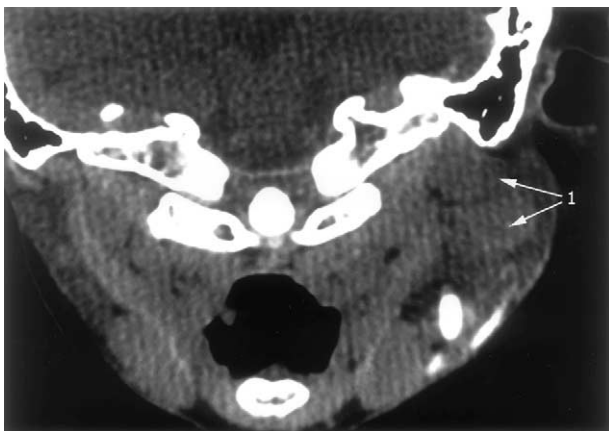
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microscopic description as well as the optimum treatment are poorly codified [3].

We report a new case of myoepithelial carcinoma discovered in an 8-year-old child. To our knowledge, this uncommon tumour has never been reported in this age group in the literature. This case underlines the problems concerning the management of tumours of the parotid region in children and the surgical difficulties involved when an extemporaneous diagnosis is not available.

2. Clinical case

An 8-year-old girl was referred to our clinic for a non-inflammatory, slightly tender left parotid mass, which had been discovered 6 weeks before. The mass, located behind the ascending mandibular branch, was bi-lobular, deep, slightly mobile and firm. Facial movements were normal and there was no satellite cervical or facial lymph node enlargement. Examination of the external auditory canal, mouth, pharynx and overlying facial and neck skin was normal. Computed tomography revealed two intra-parotid tissue formations measuring 15 and 17 mm (Fig. 1). Needle biopsy and various serologic assays were non-contributory. After obtaining informed consent explaining the principles of parotidectomy and the surgical risks for facial movements, surgery was performed. Using a classic parotidectomy approach, we discovered a tumour in the deep parotid lobe invading the facial nerve a few millimetres after its exit from the stylo-mastoid foramen (SMF). The trunk and nerve bifurcations were impossible to recognise since they were surrounded by the tumour. Every individualised branch beyond the lesion was impossible to dissect from the well-limited, bi-
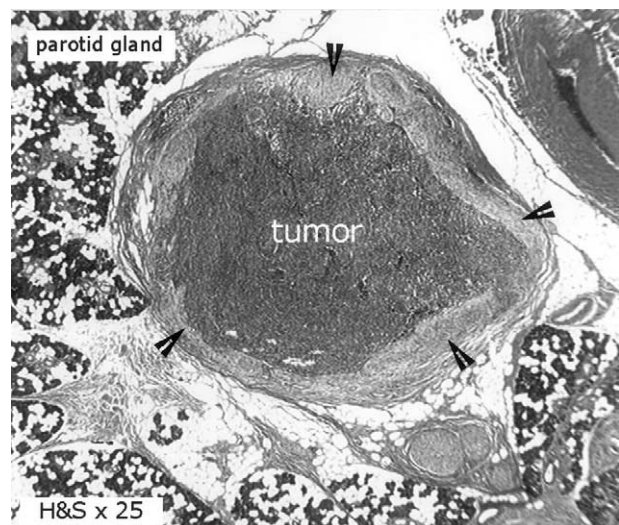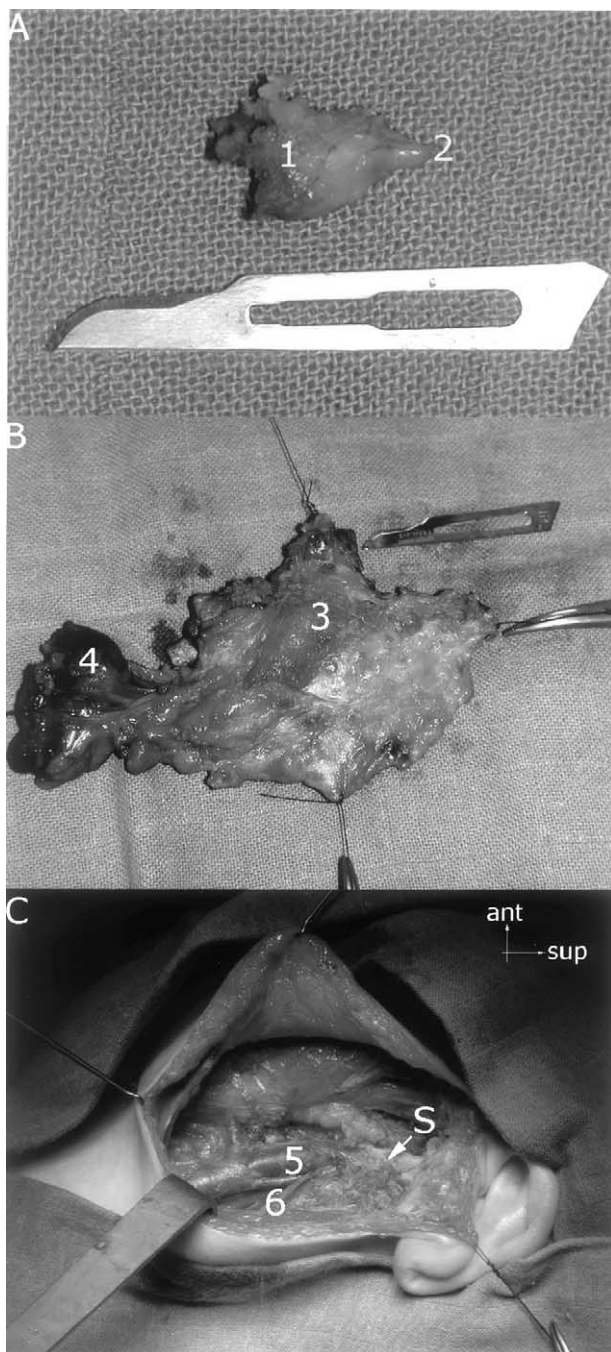
lobed mass. Macroscopically, it resembled a fairly well limited, dark red lymph node easily separated from the parotid salivary parenchyma. We performed partial enucleation for frozen-section. The pathologist's report described a soft-tissue tumour containing a few mitoses and fusiform cells but was unable to tell whether it was malignant; however, the appearance was not compatible with a lymphoma. Subsequently, we performed total enucleation requiring section of the facial nerve between its main trunk to the area where its branches exited from the parotid gland followed by a microsurgical nerve graft using a segment of the great auricular nerve between the trunk of the SMF and the temporo-facial branch.

The histologic diagnosis was difficult to make. Under light microscopy, the tumour proliferation contained basophilic fusiform cells with oval, elongated nuclei and a few mitoses. There were some areas of osteoid metaplasia. The mass was formally considered invasive since it completely surrounded the facial nerve (Fig. 2). Epithelial cell markers (cytokeratin, epithelial membrane antigen for smooth muscle actin (EMA) and S-100 protein) were positive whereas anti-desmine antibodies were negative. The tumour fixed anti-vimentine antibodies without the usual translocation seen in synovialosarcoma (X;18). These characteristics were consistent with myoepithelial carcinoma with fusiform cells and bone metaplasia.

Reintervention, involving enlarged parotidectomy with removal of the nerve graft anastomosis, revealed a 1-cm tumour nodule infiltrating the facial nerve in the SMF. Complete removal required mastoidectomy with resection of the intra-mastoid portion of the VIIth nerve and the fibrous tissue

Fig. 1 Coronal CT scan of the parotid gland showing a left tissular, bilobed intraglandular mass (1).


Fig. 2 Photomicrograph showing the tumour inside the facial nerve (arrow).

Fig. 3 (A) Tumour nodule (1) in the SMF infiltrating the facial nerve sectioned in a non-involved zone (2). (B) The parotid gland (3) removed en bloc with lymph node curage (4). (C) Petrectomy of the point of the mastoid with resection of the intra-mastoid portion of the facial nerve, tissue surrounding the SMF and extended to the styloid muscles. (5) Internal jugular vein; (6) spinal nerve; S1 styloid process.

surrounding the SMF up to the jugular foramen. The surgical specimen and curage of lymph node areas 3, 4 and 5 were removed en bloc (Fig. 3). None of the lymph nodes were invaded.
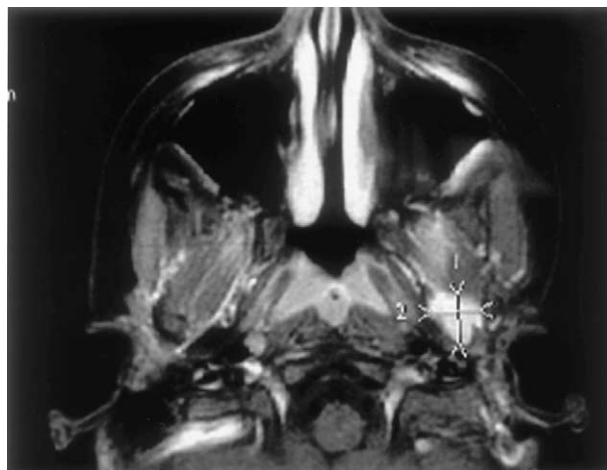
Fig. 4 Coronal (A) and axial (B) MRI scan of the parapharyngeal space showing a left tissular mass.

Eight months after this second intervention, a follow-up MRI revealed a 17 mm tumour nodule located in the pharyngeal prolongation of the parotid (Fig. 4). The subsequent, third surgical intervention, using the lateral approach, involved resection of the nodule which was in close contact to the pterygoid muscles and the ascending branch and condyle of the mandible. Sub-periostal dissection allowed enlarged removal. Histologic examination revealed that the nodule was well circumscribed by a fibrous capsule and that the tumour proliferation closely resembled the one previously described. The tumour both surrounded and infiltrated the mandibular nerve.

Eighteen months after this third surgical intervention, follow-up MRI has shown no evidence of tumour recurrence or lymph node involvement. The child has complete, global facial nerve palsy with anaesthesia in the territory of the mandibular nerve. If tumour remission persists, rehabilitation therapy for the facial palsy will be scheduled.

3. Discussion

Parotid tumours are rarely discovered in children. The classic differential diagnoses are acute or chronic cervical lymphadenopathy, vascular malformations and persistent remains of the first brachial cleft. In our experience of 158 parotidectomies over the last 5 years, only 14 concerned children (9%). They consisted in five cases of intra-parotid adenitis (two due to atypical mycobacterium), three pleomorphic adenomas, three fistulas of the first cleft, two cystic lymphangiomas and one case of myoepithelial carcinoma. According to Bull [4], only 2.5% of all malignant tumours of the

neck and face in children occur in the parotid region. For Hicks [5], only 5% of malignant salivary gland tumours occur in this age group. Nevertheless, tumours located in the parotid region have a fairly poor prognosis in children since, according to Triglia [6], they are malignant in one out of three cases compared with one out of five in adults. In his series of 261 cases, Shuller [7] found that 57% of the tumours were malignant when the mass was firm in consistency. Other factors suggesting a poor prognosis are facial palsy, pain, rapid tumour growth and the presence of ipsilateral lymph node enlargement in the cervical region. In contrast, Bluestone [8] noted that an ordinary-appearing clinical presentation could be falsely reassuring.

While X-ray studies remain indispensable, they are not very useful for establishing an accurate preoperative diagnosis. Ultrasonography can be helpful for determining the topography and dimensions of superficial tumours. Computed tomography and MRI also provide additional valuable information, especially with respect to deeper structures, particularly in the parapharyngeal area. Facial nerve infiltration can be detected by abnormal enhancement of a nerve segment and increase in nerve diameter during MRI [9]. When these two signs are present, the risk of peroperative ablation of the facial nerve increases. In our case, needle biopsy of the tumour was not helpful for the diagnosis.

We encountered particular problems in this case because this tumour is rare and difficult to diagnose, there was massive although infra-clinic invasion of the facial nerve, and the mass was multi-nodular. Further difficulties were the fact that the tumour occurred in a very young child and that the facial nerve had to be sacrificed. The diagnosis of myoepithelial carcinoma is exceedingly difficult because pathologists do not routinely encounter myoepithelial cells, making confirmation of malignancy arduous. Myoepithelial cells accelerate saliva excretion and are located between the glandular epithelium and the basal membrane of the acini of intercalar and striated canals [10]. Five tumour types have been individualised according to their cytology: clear, epitheloid, plasmocytoid, fusiform or mixed. The fusiform type found in this case report could suggest other diagnoses. The condensed appearance of the tumour and the absence of glandular elements was compatible with sarcoma according to the different types which have been described in the parotid gland [11]: leiomyosarcoma, fibrosarcoma, synovialosarcoma, malignant schwannoma. However, unlike sarcomas, myoepithelial carcinoma usually fixes epithelial markers (cytokeratin and epithelial membrane antigen) during immuno-histologic study. Other markers are present in varying degrees [3]: vimentine (100%), S-100 protein (100%), calponin (75%), smooth muscle actin (50%), gliofibrillary protein acid (31%). The majority of myoepithelial carcinomas develop in a pleomorphic adenoma [1,3,12]. When that is the case, the carcinomas are low-grade [11]. Among 25 cases, Savera described ten isolated tumours, two of which appeared in a benign pleomorphic adenoma and 15 developed in a pleomorphic adenoma [3]. When they appear in isolated form or de novo, as in our case, the carcinoma is often a high-grade malignancy [13].

When a malignant epithelial tumour is discovered, total parotidectomy is mandatory [14]. In the case we present, parotidectomy was delayed until reintervention, once the histologic diagnosis was definitely established. The facial nerve should be spared if it is macroscopically free of infiltration [15]. Thus, in children, dissection sparing the facial nerve can be performed without increasing the incidence of recurrence when the tumour is a muco-epidermoid carcinoma [4] or contains acinous cells [16]. However, Spiro considers that the nerve should be removed if invasion is seen peroperatively, whatever the cell type [17]. In two myoepithelial carcinoma series reported by Savera [3] (15 cases) and Nagao [18] (seven cases), no particular strategy is recommended even though, in four of Nagao's cases [18], there was histologic perineural invasion. This finding argues against sparing the facial nerve. In our case, conservation of the facial nerve was impossible during the first operating time; subsequent microscopic study clearly supported this decision. Nevertheless, before surgery, facial movements were clinically normal. Unfortunately, the need to remove the end-to-end nerve microanastomosis obviated the possibility of postoperative rehabilitation. Massive nerve infiltration, confirmed by the persistence of tumour remains in the SMF, forced us to extend resection to the mastoid portion of the VIIth nerve. The third intervention, showing infiltration of the mandibular nerve by the nodule's parotid pharyngeal extension, confirmed the tumour's tropism for nerve tissue. We found no lymph node involvement. This complication is reported to be unusual [3], but we feel that it is reasonable or even indispensable to perform ipsilateral functional curage.

Adjuvant chemotherapy and radiotherapy have not been found helpful for treating these tumours. In spite of the observed nerve infiltration which suggests the presence of a high-grade malignancy,

we feel that the absence of vascular emboli, of lymph node metastasis and the small number of mitoses suggested that the tumour was only weakly aggressive. Along with Lack [14], we consider that extended surgical ablation alone avoids radiotherapy and its complications in children: trismus, retarded growth of the facial bones, pituitary insufficiency and X-ray induced cancer [19]. Indeed, among seven of Nagao's cases receiving complementary radiotherapy after surgery, there were three remissions and four deaths.

Prolonged clinical and radiological follow-up is necessary. Di Palma [13] reports a patient who died from this disorder 35 years after his tumour was detected. The risk of local recurrence is high. Ten out of 18 patients in Savera's series [3] had local recurrence and two of nine followed by Nagao [13] shared the same fate, representing a 44% recurrent rate if the two series are combined. MRI of the neck and face, which can detect infraclinic tumours and cervical lymph node disease, is particularly useful for follow-up. Visceral or lymph node metastasis (to the lungs, brain, bone, skin and cervical lymph nodes) were seen in 47% of the patients followed by Savera [3]. Among 17 patients in this series who have appropriate follow-up, 59% are still alive after 42 months without any sign of recurrence, 29% have died from the disorder within 32 months and two are still alive but have metastasis. According to Nagao [18], there is no correlation between the histologic and clinical presentation since three patients who had atypical cells with numerous mitoses were cured whereas other patients who had a tumour which was histologically described as low-grade had early metastases. In the light of these findings, it is certainly reasonable to remain very cautious.

Acknowledgements

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