



Primary Synovial Sarcoma of the Inner Ear: A Case Report

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Abstract

Ear synovial sarcoma is thus rare, to our knowledge, there has been only one reported case in English medical literature. Here we report a second case of a locally advanced synovial sarcoma affecting the inner ear in a thirty-year-old female managed by chemotherapy and radiotherapy. The objective of the present paper was to describe and discuss the diagnosis and treatment of this rare malignant inner-ear tumor.

Introduction

Malignant tumor of the ear is infrequent, with an incidence of 1 per six million of the population. Whatever the histological type, an inner ear location is exceptional¹. Diagnosis should not be delayed, so as to allow satisfactory tumor removal without risk of neurologic sequelae.

Case Report

A 30-year-old female presented in the medical oncology department of national institute of oncology with a painful contracture of facial muscles with a six months history of otorrhea associated with intermittent otalgia. Physical examination revealed a budding tumor filling the external auditory canal associated with complete peripheral right facial nerve palsy. Computed tomography scan of the face was performed and confirmed the presence of a 56*37 mass extending to the tympanum and external auditory canal with osteolysis of the mastoid [figure 1,2] , there was no evidence of metastasis to lung or other sites.

A biopsy was performed and the diagnosis of a monophasic synovial sarcoma FNCLCC grade II was rendered .

Histological analysis of the tumor revealed a massive infiltration by an elongated cells with spindle shaped nuclei. The reticulin pattern was heavy with prominent vascularity and some dilated vascular channels. There were several foci of distinct cartilaginous differentiation within the tumour. Immunohistochemical study revealed a positive reaction for EMA and PSA 100 but

negative for CD34, HMB45, KL1 and desmine. A final diagnosis of monophasic synovial sarcoma FNCLCC grade II was made.

Surgical approach wasn't possible because of the extending of the tumor and the patient underwent three cycles of neo adjuvant chemotherapy consisting of doxorubicin (60mg/m²day1) and ifosfamide (3g/m² day 1 to 3) regimen.

CT scan after final courses showed a progressive disease [figure 3] with skin ulceration, local bleeding and lung metastasis. A palliative radiotherapy was indicated followed by oral

Cyclophosphamide. The patient died after 6 months.

Discussion

Although rare, synovial sarcoma is one of the most common malignant soft-tissue sarcomas in children and adolescents, the head and neck is involved in 6.8% of all synovial sarcomas².

Ear location is extremely rare , only one case of a primary synovial sarcoma of the middle ear has been reported by O'keefe et al³ in 1993 .

Synovial sarcomas often arise in areas remote from structures containing synovial membrane such as joints or bursae. Mackenzie et al suggested that synovial sarcomas arise from undifferentiated mesenchymal tissue which retains the potential for synovioblastic differentiation . Immunohistochemical studies by Abenoza et al support the theory of mesenchymal origin demonstrating a positive immunoreaction for the epithelial markers, epithelial membrane antigen [EMA] and cytokeratin [CK]

The tumour is classically biphasic, containing both epithelial and spindle cells. The biphasic nature of the tumour is the only diagnostic histological criterion although a monophasic variety where either cell type predominates is recognized as in this case.

Treatment of ear tumor depends on the degree of extension [4]. Classically, it associates a complete surgical resection of the primary tumor and/or radiochemotherapy. Surgery should follow the rules of cancer surgery rather than the general principles of ear surgery [5].

removal is incomplete in 50% of cases, accounting for the high rate of local recurrence at 10 months [4]. Prognosis is generally poor but depends on the time to

diagnosis [6].

Conclusion

Inner-ear synovial sarcoma is a rare malignant tumor that requires early diagnosis to allow a complete surgical resection which is the mainstay of Treatment

A multidisciplinary approach associating otologist ,surgeons,radiologist and oncologist is essential for efficient management.

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Illustrations

Illustration 1

Axial bone marrow reconstruction on CT scan showing an osteolysis of the temporo mandibular articulation

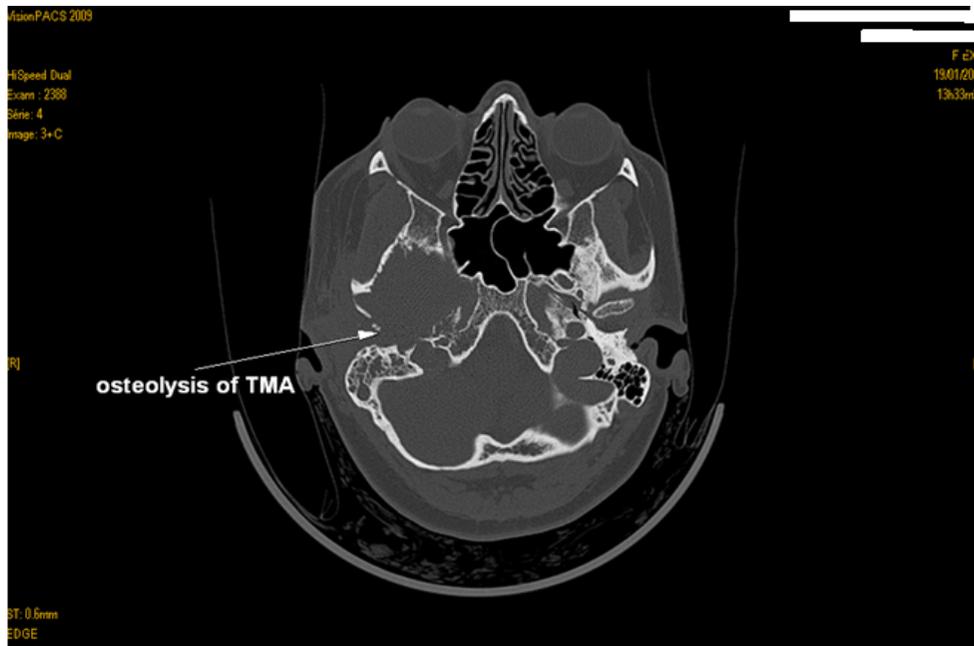


Illustration 2

Axial parenchymal window on CT scan showing a tissular process sitting in the right temporal fossa

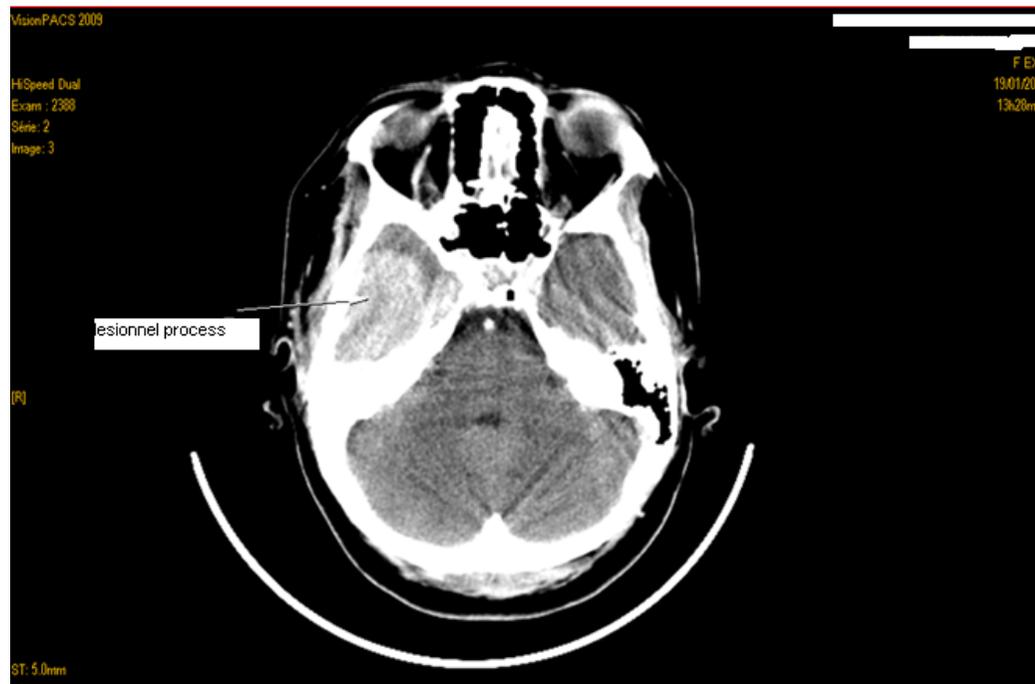
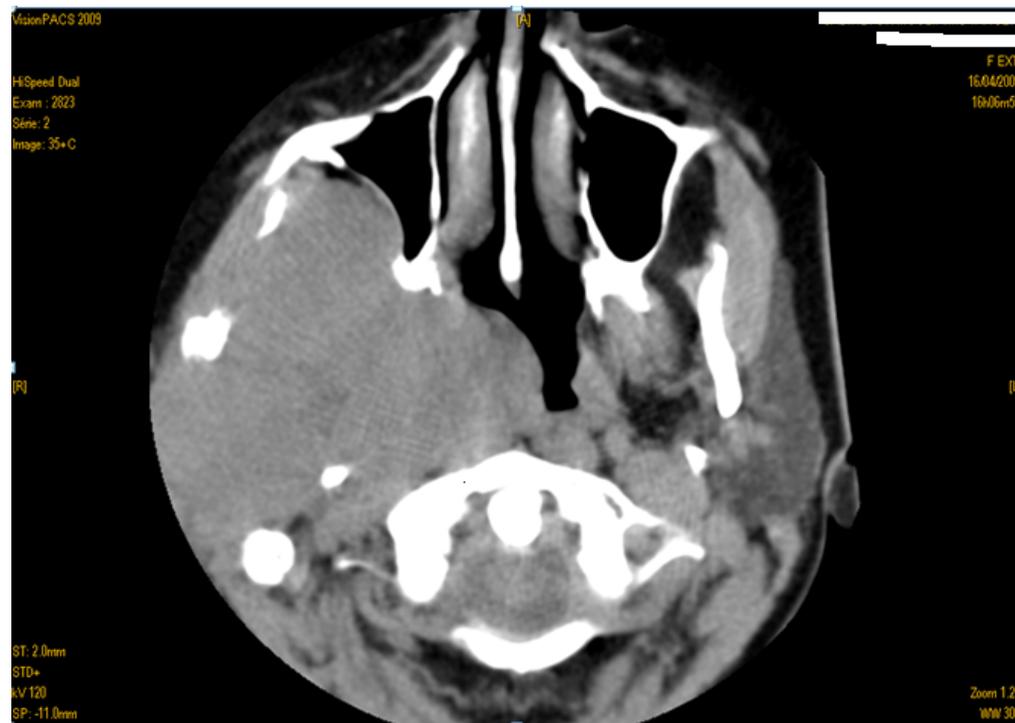


Illustration 3

Axial parenchymal window through the paranasal sinuses showing an increase of the lesion with a mass effect of the nasopharynx and osteolysis of



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