Occipito-Cervical Extracranial Schwannoma Transforming Into A Malignant Peripheral Nerve Sheath Tumour (MPNST)

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Abstract : Benign solitary schwannomas arise often from cranial nerves, but they may be located at any part of the body, especially in the face and neck region. A 27-year old woman was accepted in our clinic for an extracranially growing tumour at the occipito-cervical region. She had been operated twice in another hospital for this tumour before she was admitted to our clinic. The patient was reevaluated and the tumour was extirpated. The histopathologic examination was confirmed the previous diagnosis of shwannoma. But ten months later the patient was reoperated for a growing tumoral mass at the same place. After radical resection pathologic examination revealed malignant transformation from schawannoma to neurogenic sarcoma. Then the radiotherapy was planned, so 3500 rad was applied and the patient was seen 5 years later.

In schwannoma, malignant transformation and role of radiotherapy combined with radical resection on its treatement were discussed.

Key Words : Benign schwannoma, malignant periperal nerve sheath tumour, peripheral nerve tumours.

ITRODUCTION

The tumours of peripheral nerve sheath are rare. Among them, the benign solitary schwannoma can occur in the body elsewhere (1). But it is seen commonly at the head and neck region (1,2,6,7,). Malign schwannomas are commonly associated with neurofibromatosis and may be seen either in patients who has undergone radiotherapy or solely as a solitary tumour (6).

CASE REPORT

27-year old female patient has been complaining of a mass lesion on her neck for 8 years. The lesion was gradually enlarging and was operated twice with an interval period of three years, consequtively, five and two years ago. On physical examination there was a mass lesion which was located on the occipito-cervical region and covered with epidermis, reddish coloured and 8 cm diameter (Fig. 1). There was no sign of the Von Recklinghausen's disease and the neurological examination was normal. Routine blood biochemistry and radiologic investigations were all normal. The patient was operated and the capsular tumour attached to the occipital periosteum and musculus semispinalis capitis was removed totally with his attachements. The histological diagnosis was neurinoma (Fig.2). 10 months later the lesion was remarkably enlarged reaching a mass of 4x3x2 cm. The patient was reoperated. At the reoperation it was seen that the tumour eroded the occipital bone. This region was cauterized and the tumour was removed totally with the peripheral attachments.The pathologic diagnosis was neurogenic sarcoma (Fig.3). The petient was underwent radiotherapy and she has no recurrence up to now .

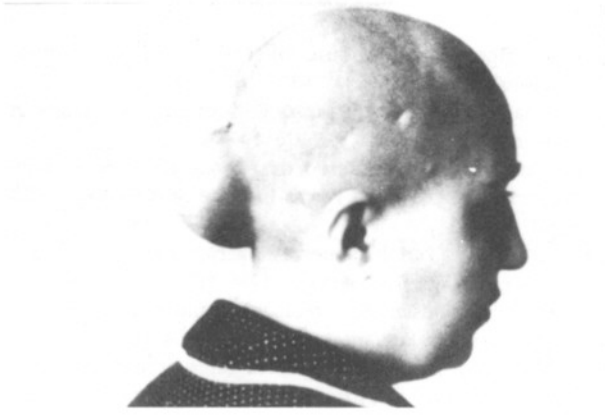

Fig. 1: The mass lesion at the occipito-cervical location.

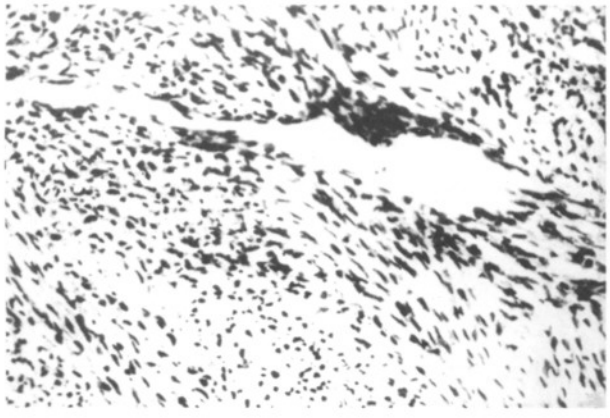

Fig. 2: Microscopical appearence of the benign schwannoma. HEx100. Uniform and ovoidal cells were seen inside of the thick capsule. There is no pleomorphism and atypical cells.

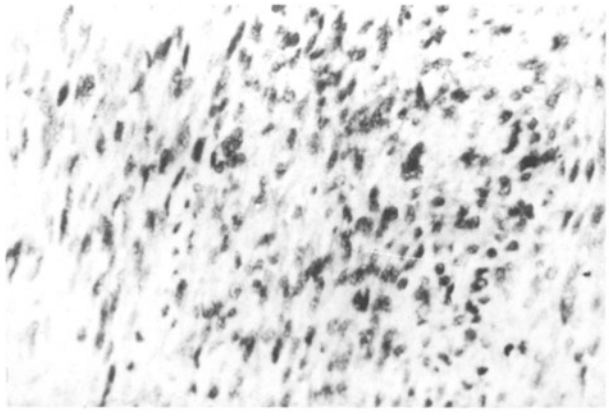

Fig. 3: Microscopical appearence of the tumour after the malignant transformation. HEx200. The tumour is circumscribed by a thin fibrous capsule and hypercellularity, pleomorphism and increased mitotic activity are seen. The invasion of a vessel wall with the tumoural cells is seen.

DISCUSSION:

The incidence of the benign solitary schwannomas located on the head and neck region is 45% and commonly seen at the lateral region of the neck (1,6,7). They are slowly growing tumours and at follow - up admissions, may reach to collosal dimensions. (1,6).

The treatment is the surgical excision and they rarely show malign transformation (2,4). According to the localization of the tumour, the reported patient is similar to the literature, but the malignant transformation is not. The solitary malign schwannomas orginate from the minor nervous elements e.g. cutaneous nerves, generally (1,7). There is no predilection in sex, and age. While Das Gupta (1) declines that the malignant schwannomas are seen at the 3rd and 5th decades in generally, Youmans (7) published that the benign schwannomas are seen before the 20 yeears old. Malignant schwannomas are immobile and can be differentiated from the benign schwannomas only histologically (1,2).

The differentral diagnosis should include differentiated fibrosarcoma,and leiomyosarcoma (3). Thet reatment of choice is the surgical excision (1,4,6,7). Occasionally, recurrences or malignant transformations were published (2,3). The treatment of malignant schwannoma is radical surgical excision of the tumour with the peripherally attached tissues,otherwise the incidence of local recurrences reach up to 40 - 73 % because of the pseudoencapsulation of these tumours.(2,6,7). Malignant schwannomas can expand along the nerve sheath but distant metastases are very rare (7). As malignant schwannomas are radioresistant,radiotherapy isn't effective alone but after the surgical resection,3000 - 9000 rad could be applied (1,7). There is no role of chemotherapy in the treatment of malignant schwannoma (1).

There were twice recurrences of the tumour as being a benign lesion transformation and on the 4th time it showed malignant since the last surgical excision there is no recurrence up to now.

It is known that the malignant transformation can commonly be seen patients with Von Recklinghausen's disease and the signs of this disease must be investigated in patients with benign schwannoma both clinically and histologically.

In conclusion, the treatment of choice of benign solitary schwannomas should be surgical excision including its capsule. Where as the malignant solitary schwannoma should be removed totally with the peripheral tissuses and radiotherapy should also be included in the tretment regime.

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