CASE REPORT

Subcutaneous Metastasis of Olfactory Neuroblastoma – An Uncommon Presentation

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SUMMARY

Olfactory neuroblastoma is a rare, slow growing, malignant tumour of neuroectodermal origin that begins in neuroepithelial cells of the olfactory membrane. A metastatic rate of 20% to 60% is reported with the most common site being the cervical lymph node. Other sites include the parotid glands, skin, lungs, bone, liver, orbit, spinal cord and spinal canal. We describe a case of olfactory neuroblastoma presented to us with scalp metastasis.

KEY WORDS:
Scalp metastasis, Olfactory neuroblastoma, Esthesioneuroblastoma

INTRODUCTION

Olfactory neuroblastoma is a rare, slow growing, malignant tumour of neuroectodermal origin that begins in neuroepithelial cells of the olfactory membrane. It was first described by Berger[1] in 1924, and since then approximately 300 cases have been reported[2]. The incidence curve for this disease has a bimodal shape with the first peak in the 2nd decade and 2nd peak in 6th decade. The clinical course is characterized by local aggressiveness and less commonly distant metastasis.

CASE REPORT

A 56 year old male presented with two scalp swellings for 2 months duration. There was no history of fever, or trauma. He was diagnosed to have olfactory neuroblastoma 4 years ago (Kadish stage A: $T_1N_0M_0$). The tumour involved the nasal cavity and sparing paranasal sinuses and he had undergone excision of the tumor and followed by radiotherapy. He was on regular follow up and there was no sign and symptom of local recurrence such as progressive nasal obstruction, epitaxis, facial pain and watering of the eyes until current presentation.

On examination, there were two smooth, firm, mild tender scalp swelling which measured 2x4cm and 3x4 cm respectively. There were no skin changes. The rest of the scalp was essentially normal. On palpation of the neck, no lymph nodes were found enlarged. Systemic examination also did not reveal any abnormality. On investigation, routine blood and urine tests for cathecolamines were within normal limits. CT scan of brain and scalp (Figure 1) showed a soft tissue mass arising from the scalp without any skull or intracranial extension. Fine needle aspiration cytology (FNAC) of the mass and biopsy was undertaken. The histopathological examination showed small tumour cells with glial fibrillary background (Figure 2a). The cells are positive for glial fibrillary acidic protein (GFAP) (Figure 2b) and neuroendocrine markers such as synaptophysin (Figure 2c). Hence, this was reported as metastatic olfactory neuroblastoma. He was then referred to oncology unit for chemotherapy and radiotherapy. There was no other distant metastasis as evaluated by CT scan of thorax, abdomen and pelvis. The patient was started on a Cisplatin-based regimen and on radiotherapy. On subsequent follow up, the subcutaneous swelling subsided and he is currently under regular follow up by an oncologist and otolaryngologist.

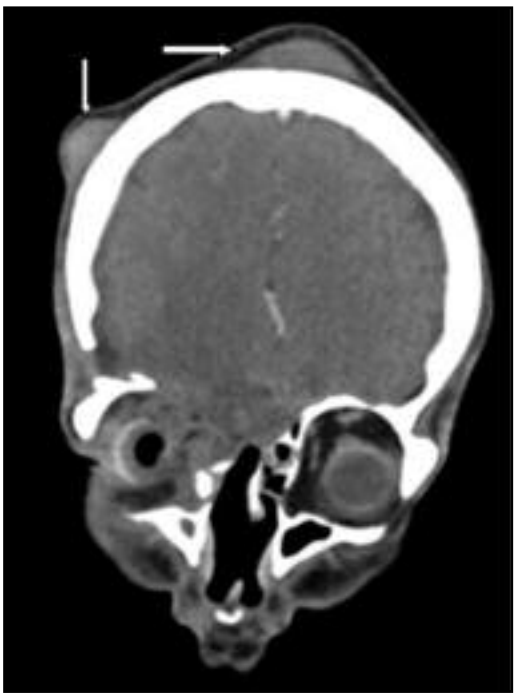

Fig. 1: Coronal view of CT scan showed two scalp swellings (arrow).

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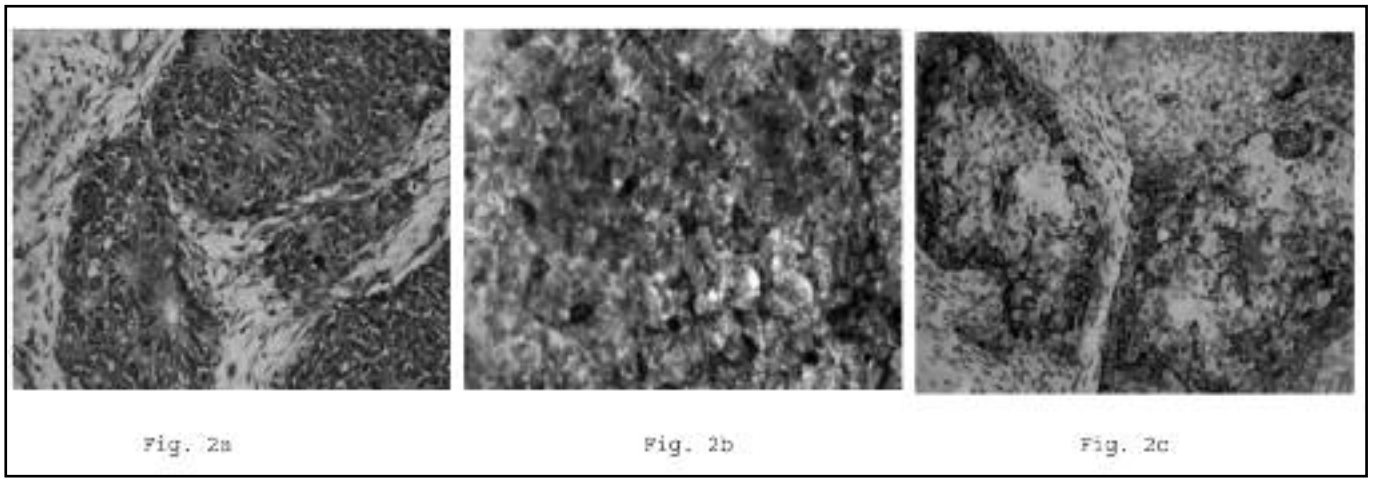

Fig. 2a : Round to ovoid tumour cells, arranged in clusters against a background of loose connective tissue. Tumour cells exhibited mild to moderately pleomorphic nucleus and small inconspicuous nucleolus, with minimal to moderate pinkish cytoplasm. Both Homer-Wright and Flexner-Wintersteiner rosettes are seen.(arrow)
Fig. 2b : Tumour cells expressing glial fibrillary acidic protein (GFAP).
Fig. 2c : Tumour cells expressing synaptophysin.

DISCUSSION

Olfactory neuroblastoma is locally aggressive and cause metastasis by lymphatic and hematogenous routes. Local recurrence has been reported in up to 57% of patients. A metastatic rate of 20% to 60% is reported with the most common site being the cervical lymph node. Other sites include the parotid glands, skin, lungs, bone, liver, orbit, spinal cord and spinal canal[3].

Metastasis to the central nervous system is infrequent and usually identified only on post-mortem examination. There are reported cases of metastasis to the adrenal glands, face, aorta, spleen and ovary[4]. However, there was no reported case of isolated scalp metastasis with no facial involvement prior to this case.

No definitive consensus regarding the optimal treatment has been reached and considerable controversy exists over its optimal management, because of the rarity of this malignancy. Multimodal therapy has been shown to improve survival in these patients. Surgical resection is the treatment of choice, particularly for locally contained low-grade tumours. Neoadjuvant radiotherapy appears to be helpful. Chemotherapy with cisplatin based regimens is helpful for high-grade malignancies.

The main prognostic factors for esthesioneuroblastoma are Kadish stage and positive lymph nodes. The overall 5-year survival rate for esthesioneuroblastoma were ranging from 45% to 64% with regional relapse rate of 26% and distant metastasis rate of 19%[5]. Although this patient initially presented with Kadish A disease with negative lymph nodes and treated aggressively with surgery and radiotherapy, he still presented with scalp metastasis. Therefore, even cases with good prognostic factors, comprehensive monitoring and follow up of patients with esthesioneuroblastoma is crucial in early detection of regional relapse and distant metastasis.

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