HEAD AND NECK TUMOURS IN CHILDREN - A REPORT ON THREE CASES

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Introduction

Tumours of the head and neck region are interesting clinical entities, although they are not very common in childhood. There are certain tumours of the head and neck region, which occur most commonly in childhood; e.g. Histiocytosis X, rhabdomyosarcoma, angiofibroma, neuroblastoma etc. The aetiology of these conditions is unknown or they are of embryonal origin. There may be single system involvement or multisystem involvement. Treatment consists of surgery, radiotherapy or chemotherapy either singly or in combination. Spontaneous resolution of some of these tumours has also been reported. One case each of histiocytosis X of frontoethmoid region, rhabdomyosarcoma of temporal bone and recurrent juvenile nasopharyngeal angiofibroma in paediatric age group which were successfully managed are reported.

Case Reports

Case 1

A 3-year-old male child presented with unilateral, progressive, painless proptosis of the right eye of one month duration (Fig 1). He had no other ocular or constitutional symptoms. There was no history of trauma to the eye. Examination of right eye confirmed axial proptosis. Ocular movements and vision were normal. ENT examination, general and systemic examination did not reveal any other abnormality. Routine haematological and urine examination were within normal limits. Radiograph skull - lateral view showed a large lytic lesion in the right frontoethmoid region. CT scan of skull and orbit confirmed the plain radiograph finding with erosion of the anterior skull base (Fig 2). Radiograph chest was normal.

To confirm the diagnosis, excision biopsy was done by external ethmoidectomy approach under general anaesthesia. There was erosion of the anterior skull base on the right side and dura was found exposed. A jelly like mass was removed and sent for histopathological examination. No intra-cranial extension was detected. Histopathological examination of the mass confirmed Histiocytosis X. Multisystemic involvement was excluded by bone scan, bone marrow biopsy, LFT, ultrasonography of the abdomen, all of which were normal. The patient was treated with 6 cycles of chemotherapy at 3 weeks interval, each cycle consisting of Inj

Cyclophosphamide 100 mg IV on day 1 and oral Prednisolone 30 mg from day 1 to day 5. Patient was asymptomatic at 6 months follow up after surgery and 1 month after chemotherapy. A repeat CT scan of skull and orbit showed a mass lesion in the same region suggestive of recurrence. Re-exploration was done by the same approach. No tumour mass was found. Remnants of the anterior skull base were biopsied which on HPE was found to be

Fig. 1 :

Fig. 2 :

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Fig. 3 :

normal. Patient has remained asymptomatic at 1 year follow up following initial surgery.

Case 2

A 5-year-old malnourished male child presented with foul smelling sero-sanguinous discharge from right ear of 3 months duration, ipsilateral facial palsy of 2 months duration and a postaural fistula of 1 month duration. Examination showed a undernourished child who was febrile and toxic. Local examination of right ear showed forward displacement of pinna and a post aural sinus with granulation tissue protruding through it along with foul smelling discharge (Fig 3). Left ear was normal. The patient had sensorineural hearing loss and lower motor neuron facial palsy on the affected side. Differential diagnosis entertained at this stage was chronic otitis media with complication, tuberculous otitis media and rhabdomyosarcoma. Radiograph mastoids showed sclerosis of both mastoids with no erosion of the tegmen or sinus plate. Radiograph chest was normal. Exploration of the right mastoid was done under general anaesthesia using a post aural approach. Pale granulation tissue was found occupying the entire mastoid and middle ear. Tegmen and sinus plate were found intact. There was a dehiscence of the horizontal part of the fallopian canal and extension of disease medially to the petrous temporal bone through the hypotympanic cells. In view of the poor general condition of the patient extensive surgery was not undertaken. Histopathological examination of the excised tissue revealed embryonal rhabdomyosarcoma. The patients parents could not afford a CT scan which would have shown the exact extent of involvement. He was referred for further management with chemotherapy and radiotherapy.

Case 3

A 10-year-old male child, known case of recurrent juvenile nasopharyngeal angiofibroma, operated 4 times in the past 4 years presented with right sided nasal obstruction, serosanguinous nasal discharge of 3 months duration. The last surgery was done about 1 year ago. General and systemic examination did not reveal any abnormality. Examination of the nose showed a well healed Weber Fergusson incision on the right side, widening of the nasal bridge and a fleshy mass in the right nasal cavity. Posterior rhinoscopy showed a fleshy mass in the nasopharynx obstructing both choanae and extending upto the roof. Routine haematological tests, urine analysis, and radiograph chest were normal. Radiograph (lateral view) of the nasopharynx showed a soft tissue opacity in the nasopharynx. Contrast enhanced CT scan showed a mass occupy-

Fig. 4 :

Fig. 5 :

ing the right nasal cavity, nasopharynx, right maxillary sinus and right infratemporal fossa (Fig 4). The tumour was excised under GA through a combined Weber Fergusson, transpalatine and transmaxillary approach after ligation of the right external carotid artery. The lobulated tumour mass measuring 7 cm x 5 cm was sent for histopathological examination and was reported as angiofibroma (Fig 5). Post operative recovery was uneventful except for a 2mm oroantral fistula which closed spontaneously after 1 month. No recurrence was detected at 3 months follow up.

Discussion

In children, other than lymphomas, rhabdomyosarcoma, neuroblastoma, histiocytosis X, nasopharyngeal and thyroid carcinomata occur rarely. The incidence of juvenile nasopharyngeal angiofibroma is one in 15000 [1]. The aetiology of all these conditions remains obscure.

Histiocytosis X, earlier known as eosinophilic granuloma, Hand - Schuller - Christian disease and Letterer - Siwe disease was given the present name by Lichenstein in 1953 [2]. The symptoms are skin rash,

recurrent aural discharge, bone pain, lumps in scalp, proptosis, breathlessness and lymphadenopathy [3]. On radiograph, the skull lesions appear as 'punched out areas. Pulmonary and cervical lymph node involvement is common. Hepatosplenomegaly indicates poor prognosis. Electron microscopy shows 'inclusion granules known as 'Birbeck granules [4]. Single site disease is treated with intralesional steroids or radiotherapy if the lesion is deep seated. Systemic steroids are used in multicentric disease. Alternatively, chemotherapy and steroids is recommended. Currently chemotherapy and radiotherapy is preferred to surgery [5]. In our case steroids and cyclophosphamide were used, as the lesion was not suitable for intralesional steroid infiltration or radiotherapy.

About a third of rhabdomyosarcomas arise in the head and neck region. The common sites are orbit, anterior facial structures, middle ear, nasopharynx, palate, pterygopalatine region and paranasal sinuses. When the mass arises below a mucus secreting epithelium as in the present case, it has a 'botryoid or 'grape-like appearance that is diagnostic. The treatment consists of surgery with or without radiotherapy for local control, together with chemotherapy. Single site tumours have a better prognosis than multiple site tumours [6]. Involvement of the skull base or meninges is a poor prognostic indicator [7]. Hence, pre treatment assessment by CT and or MRI scanning is important [8]. The present case had erosion of the skull base in the region of the petrous temporal bone. Hence, radiotherapy and chemotherapy was advised.

Juvenile nasopharyngeal angiofibroma, a disease of adolescent males arises in the region of the sphenopalatine foramen. Lateral extension takes place through pterygo-palatine fossa, infra temporal fossa,

cheek or the orbit. Incomplete removal results in regrowth of the residual mass. In recurrences, as in the present case, where the infratemporal fossa was involved, the recommended approach is Weber-Ferguson combined with a transmaxillary - trans nasal approach [9]. Pre-operative embolisation of the feeding vessels reduces blood loss and post operative morbidity [10].

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