Isolated Hypoglossal Nerve Palsy Due To Chordoma

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Abstract : Chordomas are rare tumorus arising from notochord remnants. Most cases are encountered in adult life, but rare cases have been reported in childhood.
In this report, a six-year-old boy with a chordoma in the craniovertebral junction presenting with isolated hypoglossal nerve palsy is reported.

Key words : Chordoma, Craniovertebral junction, Hypoglossal nerve

INTRODUCTION

Chordoma is a rare tumour representing less than %1 of nervous system tumors (1,2,3,6,8). In spite of their congenital origin these neoplasms appear with the greatest frequency in the 5th decade of life and are rarely encountered in patients younger than 30 years (1,2,3,8). The predominant sites are the sacrococcygeal (50%), spheno-occipital (35%), and vertebral regions (15) (2). Clinical features primarily depend on the localisation of the tumoral mass. To our knowledge, a case of chordoma located in the craniocervical region presenting as isolated hypoglossal nerve palsy has not previously been reported in the literature.

The complaints, clinical and neuroradiological features, histopathology and therapy of this uncommon entity are presented and the related literature is reviewed.

CASE REPORT

A six-year-old boy was admitted to hospital with atrophy and lateral deviation of the tongue which had first appeared two months before and progressed without any other particular symptoms.

General paediatric examination including complete ear-nose-throat examination was normal. No evident lateral cervical lymphadenopathy was palpable. Neurological examination revealed right hypoglossal nerve palsy. Atrophy was also noted at the same site (Fig. 1). The remaining neurological examination was considered normal, with no deficits affecting the other ipsilateral cranial nerves.

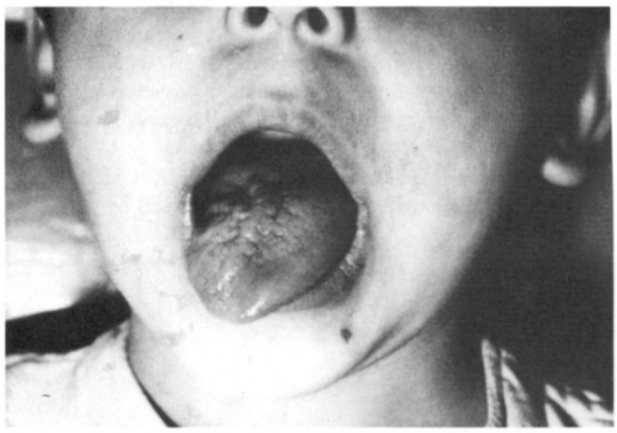

Fig. 1 : The child showing atrophy and lateral deviation of the tongue.

Plain X-ray of the craniocervical junction showed no destructive lesions. The CT-Myelogram (Fig. 2) showed a non-enhancing low-density lesion in the craniocervical junction extending from the right of the skull base to the first and second cervical vertebrae. No bony involvement was noted. Vertebral angiography revealed that the segment of the right vertebral artery, before entering the cranial cavity, was stretched and displaced by the tumoral mass.

After suboccipital craniectomy, C1 and C2 laminectomies were performed, a soft gray tumour

extending from the upper lateral cervical region to the clivus and extradural in location was seen and removed subtotally.

Histopathological examination revealed a chordoma (Fig 3). Postoperatively the patient did well. Radiotherapy was carried out with a total dose of 6000 rad over a period of two weeks.

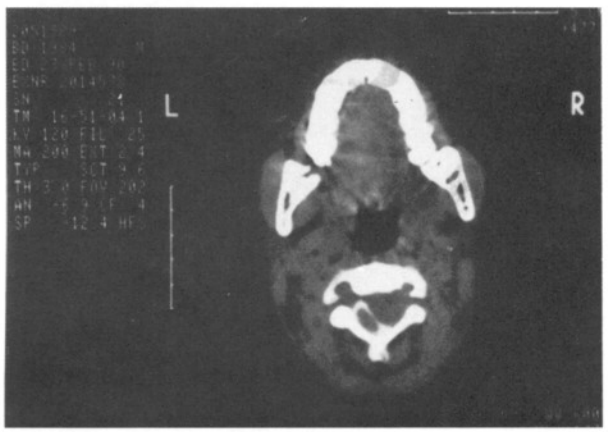

Fig. 2 : CT-Myelography of the craniovertebral junction. The spinal cord is on the left side while the tumour is on the right.

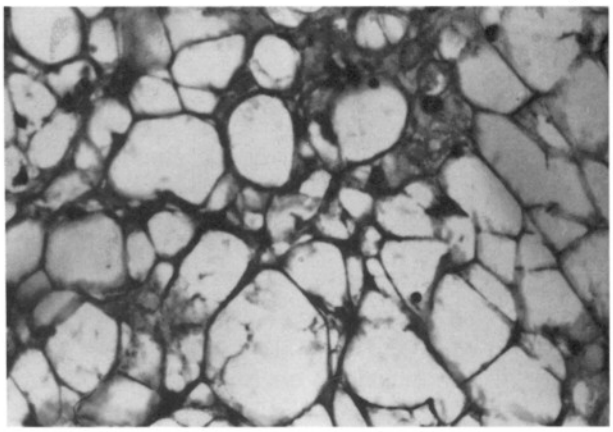

Fig. 3 : Histopathological examination showing chordoma (H.E.x460).

Follow-up examination twenty months after surgery, showed that the hypoglossal nerve palsy had improved. CT scan revealed no recurrence.

DISCUSSION

Hypoglossal nerve palsy may be due to trauma, infection, degenerative disease or tumour (4,7). Although tumoral involvement of the hypoglossal nerve has been described with lesions in the brain stem or in the base of the skull (4,5,7), isolated hypoglossal nerve palsy due to chordoma in the region of the foramen magnum has not previously been reported (1,2,3,6,8).

The clinical features of intracranial and cervical chordoma in children are similar to those in adults (1,3,6,8). They are related with compression of the neural tissue caused by the tumoral mass. Headache, neck pain, diplopia and unilateral cranial nerve palsies were the most common symptoms at presentation. In the present case, unilateral hypoglossal nerve palsy was the only prominent symptom on admission.

Generally, CT scan of cervical or intracranial chordoma shows a non-enhancing low-density lesion with or without erosion of the adjacent bony structures (1,3,6). Surgical treatment is usually successful in achieving a disease-free interval and radical surgical excision should be performed as soon as possible. Surgery plus high-dose radiation therapy has been recommended (3,6,8).

In childhood, chordoma can present with unexpected clinical findings such as diplopia or isolated hypoglossal nerve palsy as in the present case. We wish to underline that chordoma located in the region of the craniocervical junction may be a cause of 12th cranial nerve palsy. Therefore, neuropaediatricians and paediatric neurosurgeons should be take this rare tumour into consideration when dealing with a child with isolated hypoglossal nerve palsy.

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