An unusual cause of "dropped head syndrome"

Sir,

Neurofibromatosis has not been reported as a cause of "dropped head syndrome" (DHS).

A 14-year-old girl presented with weakness of upper and lower limbs of 3 months and inability to extend the neck of 1-month duration [Figure 1]. General examination revealed multiple cafe-au-lait spots and multiple cutaneous neurofibromas all over the body. There was spastic quadriparesis with decreased sensations below C3 dermatome. There was a 4×8 cm^2 right paraspinal swelling in the neck. Magnetic resonance imaging of the cervical spine showed multiple contrast enhancing intraspinal (intradural and extradural) and extraspinal lesions [Figure 2]. Dynamic X-rays of the neck were normal. Electomyography (EMG) showed denervation of cervical muscles. She underwent C1-3 laminectomy and excision of extradural and intradural neurofibromas. At surgery there was a large, vascular lesion in the intermuscular plane which was

Figure 1: Clinical picture of patient showing "dropped head"

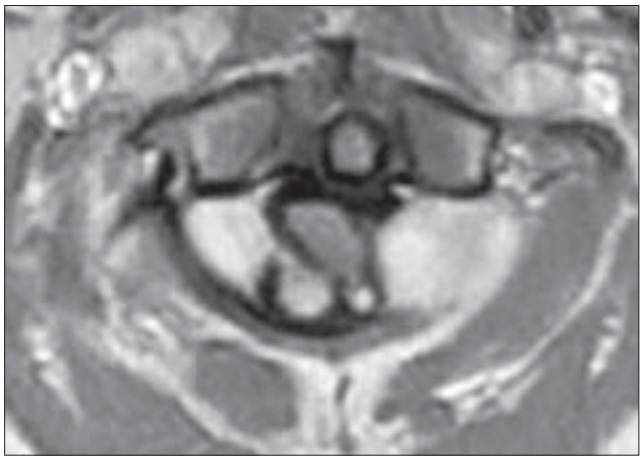

Figure 2: Magnetic resonance imaging of spine. T1W post gadolinium images in axial sections showing multiple enhancing masses in the intradural, extradural and extraspinal compartments

excised and the biopsy was reported as Malignant nerve sheath tumour (MPNST). After dural opening, there were multiple bilateral well-defined encapsulated lesions attached to roots. Three lesions, which were large and causing compression of the cord, were excised. The biopsy of these lesions was reported as neurofibroma. She was advised to continue on Philadelphia cervical collar and was referred to Radiotherapy and Physical rehabilitation for further management.

Dropped head syndrome is also known as Head drop or Head ptosis. It results from weakness of neck extensors (splenius capitis, semispinalis capitis, and erector spinae) due to various causes. Gourie-Devi *et al*, have proposed a classification for 'dropped head syndrome'.[1] They have classified DHS based on the cause into myogenic, neurogenic, miscellaneous, and local groups. There is no mention of neurofibromatosis as a cause of DHS in this classification. Various other causes like parkinsonism, syringomyelia, and anti-epileptic drugs (sodium valproate) have also been described recently.[2,3] Very few pediatric cases have been reported.[4] In our patient, bilateral multiple neurofibromas have resulted in weakness of neck extensors. In managing these patients, in addition to the specific treatment depending on the cause, special attention has to be paid toward their feeding problems and social embarrassment.[1] There is a 8-13% lifetime risk of developing MPNST's in patients with Neurofibromatosis Type I and 5-year survival rate after diagnosis is 21% *vs.* 42% in sporadic cases.[5] Neurofibromatosis needs to be considered in the differential diagnosis of a child presenting with dropped head syndrome.

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