

small mass which progressively increased in size. The patient used to walk in a flexed posture because of its weight. Neurological examination showed a conscious oriented patient with gross spasticity in the lower limbs. He had power of grade 3/5 in lower limbs and 4/5 in upper limbs, exaggerated reflexes, extensor planters and sensory level till C3

Plain X-ray of the skull showed sphenoid dysplasia [Figure 2]. Dynamic X-rays of the cervical spine showed reducible atlantoaxial dislocation (AAD). Computed tomography (CT) scan of the craniovertebral junction (CVJ) revealed AAD and rotational subluxation [Figure 3].

Magnetic resonance imaging of the cervical spine showed AAD, cord compression at CVJ and syrinx from C2 to C7 [Figure 4]. The patient was diagnosed to have NF1 with giant PNF. The atlantoaxial and atlanto

Giant plexiform neurofibroma presenting with craniovertebral junction instability

Sir,

We report a case of giant plexiform neurofibroma (PNF), presenting with progressive quadriparesis with neurofibromatosis 1 (NF1).

A 37-year-old male presented with gradually progressive spastic quadriparesis with bladder involvement since the last 1 year. The patient had a massive irregular pedunculous mass extending from the left forehead to the mid abdomen [Figure 1] with multiple neurofibromas all over the body. The mass was lobulated, firm, nontender, and it entirely engulfed the left eye, nostril and the mouth. The lesion was present since birth as a



Figure 1: Picture showing patient with large PNF



Figure 2: Plain X-ray of the skull showed sphenoid dysplasia

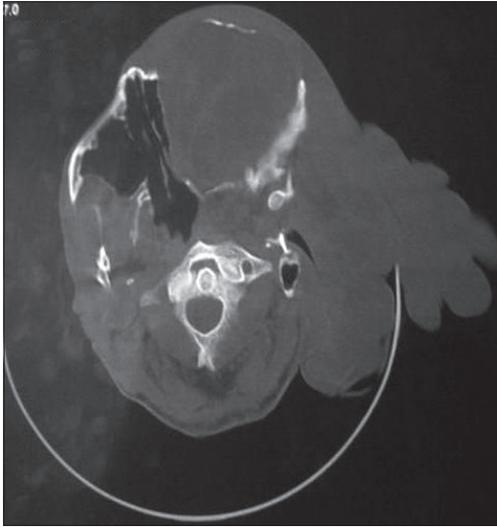


Figure 3: CT scan of the CVJ revealed AAD

occipital junction, though very stable complexes, also have an extensive mobility making them vulnerable for dislocation. Atlantoaxial instability may result from many reasons including hypoplasia of the odontoid process and from laxity of the transverse ligament.^[1] In our patient, the progressive enlargement of the PNF with subsequent increase in its weight could have led to the atlantoaxial instability. PNFs are benign tumors originating from nerve sheath cells, subcutaneous, or visceral peripheral nerves, occurring in only 5% of patients with NF1.^[2] There are case reports of atlantoaxial instability due to neurofibromatosis.^[3,4]

The patient presented at admission a Nurich's grade 3 neurological condition. A team of doctors comprising neurologists, neurosurgeon, plastic surgeon and surgical oncology counseled the patient about his condition and the patient opted for the spinal surgery. The patient underwent operation with occiput to C2 and C3 fusion (titanium loop and bone graft) and C1 arch excision. We present this case to highlight a rare manifestation of massive plexiform neurofibroma causing atlantoaxial instability leading to spastic quadriparesis.



Figure 4: Magnetic resonance imaging of the cervical spine showed AAD, cord compression at CVJ and syrinx from C2 to C7

Bindu Menon, G Samson Sujit Kumar¹

Departments of Neurology and ¹Neurosurgery, Narayana Medical College and Superspeciality Hospital, Chintareddypalem, Nellore-2, Andhra Pradesh, India

Address for correspondence:

Dr. Bindu Menon, Professor
Department of Neurology, Narayana Medical College
and Superspeciality Hospital, Chintareddypalem,
Nellore-2, Andhra Pradesh, India.
E-mail: bneuro_5@rediffmail.com

DOI: 10.4103/0976-3147.71739

References

1. Shaffrey CI, Chenelle AG, Abel MF, Menezes AH, Wiggins GC. Anatomy and physiology of congenital spinal lesions. In: Bencil EC, editor. Spine surgery: Techniques, complication avoidance, and management. 2nd ed. Amsterdam: Elsevier; 2005. p. 61-87.
2. Pivnick EK, Riccardi VM. The neurofibromatoses. In: Freedberg IM, Eisen AZ, Wolff K, Austen KF, Goldsmith LA, Kartz SI, *et al.* editors. Fitzpatrick's Dermatology in General Medicine. New York: Mc graw Hill; 1999. p.2152-8.
3. Veras LM, Castellanos J, Ramírez G, Valer A, Casamitjana J, González F. Atlanto axial instability due to neurofibromatosis: Case report. Acta Orthop Belg 2000;66:392-6.
4. Isu T, Miyasaka K, Abe H, Ito T, Iwasaki Y, Tsuru M, *et al.* Atlantoaxial dislocation associated with neurofibromatosis. Report of three cases. J Neurosurg 1983;58:451-3.