

Unusual bone formation in the anterior rim of foramen magnum: cause, effect and treatment

Atul Goel · Abhidha Shah

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Abstract A rare case of proatlax segmental abnormality resulting in a bony mass in the anterior rim of the foramen magnum is studied. Case report of a 19-year-old female showed a progressive weakness of all four limbs for about 3 years. When admitted she could not perform any useful activities by herself. Investigations revealed an unusual bone growth in the region of the anterior rim of foramen magnum that resulted in severe cord compression. The abnormal bone formation involved the lower end of clivus, the tip of the odontoid process and the posterior arch of the atlas. Dynamic imaging did not reveal any clear evidence of instability. Following transoral decompression and posterior fixation, the patient showed dramatic and lasting clinical recovery. Conclusions were drawn as follows. Anomalies of the most caudal part of the occipital sclerotomes due to the failure of proatlax segmentation can be the cause of an abnormal bone mass in the anterior rim of foramen magnum. Transoral decompression, followed by posterior atlantoaxial fixation, results in neurological recovery and provides lasting cure from the problem.

Keywords Proatlax · Segmentation anomaly · Transoral decompression · Posterior fixation

Introduction

Bone anomalies related to failure of segmentation of the most caudal occipital sclerotomes are rare, but have been

identified. The anomaly leads to abnormal bone formation in the region of anterior rim of foramen magnum, either in the midline and/or laterally. The anomaly has been occasionally referred to as ‘third condyle’, ‘remnants of occipital vertebra’, ‘un-formed bone masses on the anterior rim of foramen magnum’ and ‘prebasi-occipital arch in combination with an odontoid bone’. This anomaly has been grouped under the term ‘proatlax segmentation abnormalities’. Due to the extreme rarity of such cases, the criterion for identification of the anomaly and the management protocol have not been elaborately discussed in the literature. We describe a case and discuss the available literature on the subject.

Case report

A 19-year-old female had traumatic neck injury when she fell on the back of her head while playing 3 years before presentation for treatment in our department. Following this fall, she developed weakness of the right-sided limbs and urinary retention. She gradually improved completely in all her symptoms in about 4 months. Three months prior to her present admission, she again had an accidental fall while walking. Following this, she developed quadriparesis. The weakness now progressed. When admitted she was bed-ridden and could not perform any useful life activities by her own self. She also had urinary retention for which she was catheterized. Clinical examination revealed spasticity in all four limbs. The power in the right-sided limbs was Grade 2/5 and that in the left-sided limbs Grade 4/5. There were no definite sensory deficits. Investigations revealed an abnormal bone growth in the inferior end of the clivus that extended laterally. The tip of the odontoid process also had a posteriorly curving bone growth. The

A. Goel (✉) · A. Shah
Department of Neurosurgery,
King Edward VII Memorial Hospital,
Seth G.S. Medical College, Parel, Mumbai 400012, India
e-mail: atulgoel62@hotmail.com

posterior arch of the atlas was not identified and was probably assimilated. The abnormal clival and odontoid bone and the posteriorly buckled tectorial membrane resulted in severe cord compression (Fig. 1). Transoral decompression that involved resection of the inferior end of the clivus and the abnormal bone of the odontoid process were carried out. The abnormal bone was relatively soft. The patient was then turned prone and a posterior plate and screw fixation of the C1 lateral mass and C2 pars was carried out [2, 3]. Following surgery, the patient showed dramatic neurological recovery. At a follow-up of 23 months, she was able to walk unaided, power being graded to be more than Grade 4 and had rejoined her school (Fig. 2). The urinary function also had recovered.

Discussion

Although rare, proatlantal segmentation anomalies have been described. Most of the descriptions of clinical issues related to such anomalies are only in the form of isolated case reports [1, 4, 6, 8]. Menezes and Fenoy [5] recently reported their extensive experience on the subject and discussed their management issues.

A bone process from the anterior margin of the foramen magnum, called as the third condyle has been described [7]. This process is usually attached to or may form a joint with the dens and less frequently with the anterior arch of the atlas. Hayek [9] found such a bone formation regularly in lower animals. Schumacher [10] included it among the

Fig. 1 **a** CT scan reconstruction image showing the abnormal bone arising from the inferior end of the clivus and tip of the odontoid process. The posterior arch of the atlas is not identified. **b** Sagittal image of T1-weighted MRI showing the compression of the cord by abnormal bone growth. **c** T2-weighted image showing the abnormal bone and ligamentous mass and neural compression. **d** Coronal image of MRI showing the transverse extension of the abnormal bone as hypo-intense ridge at the level of arch of atlas

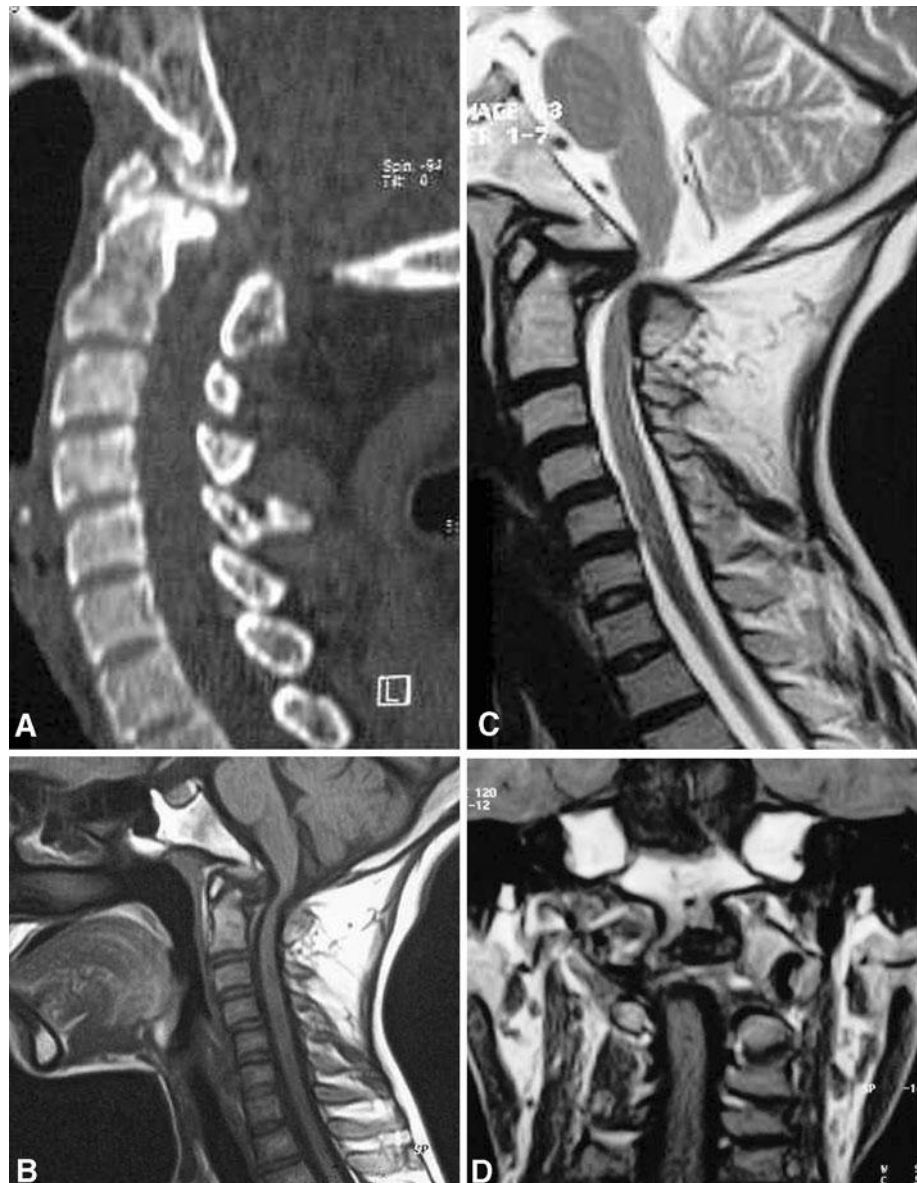




Fig. 2 **a** Postoperative CT scan showing the anterior decompression. Artifacts related to stainless steel implant are seen. **b** Sagittal image through the lateral mass showing the plate and screw fixation

manifestations of the arcus prebasi-occipitalis. It is generally in the midline, but may be paramedian. Such anomalous bone can result in neural compression and vascular compromise, and can manifest with abnormalities in cerebrospinal fluid dynamics. Menezes and Fenoy [5] identified reduction of posterior cranial fossa volume as a result of the anomaly and reported presence of Chiari malformation. The hypocentrum of the fourth occipital sclerotome (proatlas) forms the anterior tubercle of the clivus. The centrum forms the apex of the dens. The rostral ventral neural arch contributes to the formation of the

occipital condyles, third condyle and the anterior rim of the foramen magnum. The dorsal caudal part of the neural arch forms the posterior arch of atlas and the lateral atlantal masses. The persistence of anterior tubercle from the anterior margin of foramen magnum represents the hypocentrum abnormality of the proatlas. Our patient had abnormalities of the posterior arch of the atlas in addition to the ‘third condyle’ signifying a segmentation anomaly of the neural arch of the proatlas.

Most of the described patients who manifested with neurological symptoms were in their first and second decades of life. In none of the reported cases was there any clinical or radiological evidence of instability. The abnormality can be suspected when there is an abnormal bone growth in continuity with the inferior edge of the clivus. Although this was not performed in the presented case, three-dimensional CT scan can show the anomaly clearly. The bone growth may not always be large as well as neural compression may not be identified and some of these cases may be incidentally detected. Anterior transoral decompression followed by posterior fixation, as described earlier, appears to be a rationale method of treatment. Surgery results in rapid and sustained neurological recovery.

Conflict of interest statement None of the authors has any potential conflict of interest.

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