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Case Report

Plasmacytoma of Dens as a cause of Atlanto-axial instability

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A case of Plasmacytoma of dens (Odontoid Process) in an adult male managed by trans-oral excision and posterior fusion is reported for its rarity. The importance of neuro-imaging and the treatment options, in view of the associated instability at the atlanto-axial region and the difficulty in preoperative diagnosis are emphasized.

Keywords: dens; atlanto-axial; plasmacytoma; myeloma; computed tomography; magnetic resonance imaging

Introduction

Plasmacytoma of spine is not rare. However, it is uncommon for it to involve the high cervical spine[1] and as a potential cause of atlanto-axial dislocation it is seldom reported in the literature.[2] Solitary myeloma especially in the region of atlas and axis is rare. Its differentiation from tuberculosis in this part of the world is sometimes difficult[3] and often warrants a histopathological confirmation. In the context of the so called empirical antitubercular treatment, for lesions of the atlanto-axial region, without displacement and especially with intact neurology, the present case emphasizes the value of detailed neuroimaging, surgical intervention and histopathological examination.

Case report

A 35-year-old man presented with pain in the nape of the neck and occipital region for 3 months. Pain was continuous, dull aching, increased on movements of neck and relieved by bed rest. There was no history suggestive of cranial nerve dysfunction, weakness of limbs, altered sensations or disturbances of bladder or bowel function. There was no history of trauma, fever or constitutional symptoms. No personal or family history of tuberculosis was forthcoming. On examination, he was moderately built, had no anaemia or jaundice or edema or lymphadenopathy. Pulse, blood pressure, respiration and bed-side respiratory functions were normal. Budd's index was 1 : 14. Examination of the central nervous system was normal. The upper cervical spine was tender on palpation with severe paraspinal spasm restricting all movements of the neck. Abdominal examination was normal. Laboratory investigations demonstrated haemoglobin of 9.1 gm/dl, total leucocyte count of 12, 100 per cc and Erythrocyte sedimentation rate of 85 mm/1st h. Peripheral blood smear examination was normal and serum for anti-tubercular antibodies (Ig-G) was negative. He had normal renal and liver function tests. Plain X-ray of cervical spine showed loss of normal lordosis and increased prevertebral soft tissue shadow at C1–C2 with destruction of dens; alignment of spine was within normal limits. Due to severe spasm of the paraspinal muscles dynamic views could not be obtained. Computed tomography of upper cervical spine showed destruction of the odontoid (Figure 1).

Magnetic resonance imaging demonstrated replacement of dens by abnormal tissue that was hypo-intense on both T1 and T2 weighted images with increased prevertebral soft tissue. The difference in intensity was more marked on proton density images (Figure 2). There was a suggestion of involvement of the transverse ligament. With a provisional diagnosis of tuberculosis he was started on anti-tubercular treatment and a trans-oral biopsy was planned, to confirm the pathology. Pre-operatively, skull traction was applied. At trans-oral surgery, on opening the posterior pharyngeal wall a grayish, avascular fleshy mass was seen infiltrating pharyngeal constrictor muscles and the odontoid. The odontoid process was replaced by this tumour mass except for a thin shell of bone posteriorly. A near total removal of the mass was performed. The post operative period was uneventful. Histopathology of the tumour tissue revealed sheets of monomorphic round cells with scanty cytoplasm and dark nuclei infiltrating collagen and some fragments of lamellar bone (Figure 3). At some places they had plasma cell morphology.

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A postoperative computed tomogram revealed good decompression and a slight posterior displacement of C2 body. The hematological work up performed for disseminated myeloma, including bone-marrow aspiration and plasma electrophoresis was negative. In view of the suspected instability due to the extensive destruction around the atlanto-axial region a posterior Gallie's C1–C2 fusion with wire and bone was performed. The patient was mobilized with a four-post collar support and referred for radiotherapy. On follow up for 2 years, there were no features of recurrence or dissemination of myeloma.

Figure 1 Computed tomogram axial section delineates the destruction of the odontoid and there is increased prevertebral soft tissue

Figure 2 MR image sagittal tomograms demonstrate the altered signals from the region of odontoid, reaching the upper part of the axis body also, possibly involving the transverse ligament

Discussion

Myeloma ranks second only to metastatic carcinoma as a cause of paraplegia.[4] Solitary myeloma (Plasmacytoma) is diagnosed when there is evidence of only one lesion, and when aspirted marrow, is negative for neoplastic cells. This has a considerably better prognosis. Solitary myeloma should be regarded as a plasmacytoma with a capacity for bone destruction but without a tendency towards dissemination. However, the tumour eventually becomes systemic. The elevated globulin in serum is sometimes the only way of determining the dissemination and the prognosis. Bence-Jones proteinuria is present in 9% patients with apparent solitary tumours.

Cervical and upper thoracic spine are rarely affected, whereas vertebrae of lower thoracic spine are more likely to be involved (Waldenstrom).[5] Hastings *et al*[6] reviewed 12 cases where neoplasms involved the first and second cervical vertebrae. They found that lesions involve this region rarely; 3 of 12 had multiple myeloma.[6] They suggested that laminectomy could be hazardous and stabilization of the area should be considered after the needle biopsy or open trans-oral biopsy without laminectomy.

Most patients present with acute pain in the neck. At this level, the extradural spread of the tumour is

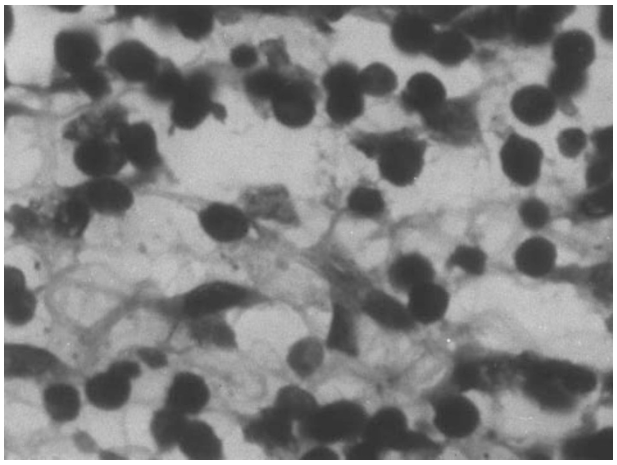

Figure 3 Microphotograph of the tumour showing monomorphic rounded cells with dark nuclei and scanty cytoplasm with fragments of lamellar bone and collagen. Cells show features of plasma cells at places $\times 10$

less likely to be the cause of cord compression. Plain radiographs are less helpful and tomograms and cine radiographs sometimes show, evidence of abnormal movement. Reduction by skeletal traction and fusion or halo-fixation would achieve spinal stabilization, and sometimes surgical intervention relieves the cord compression.[7] Mayo clinic statistics of 145 patients with myeloma disclosed involvement of C1 in four and of C2 in seven. Usually the tumour is in the articular mass of the atlas and the body of axis below odontoid. Involvement of odontoid is extremely rare and only one case has been reported in the series of Dickman *et al*[2]; while the second case had amyloidosis as a possibility along with myelomatosis. Among 53 cases where transoral approach was used, Hadley *et al* did not encounter any case of plasmactyoma and the experience of Crockard and of Menezes[8–10] was similar.

The involvement of odontoid by any lesion, singularly, is a distinct rarity. In the Indian context, tuberculosis is a common cause of acquired atlanto-axial instability.[3] However, the imaging, especially the MRI, features to distinguish the lesion from others are not described adequately. An empirical diagnosis or treatment, therefore, could be erroneous. Trans-oral excisional biopsy and stabilization, either anterior or posterior, appear to be the ideal procedures in this situation. Solitary myeloma treated by excisional biopsy and postoperative radio-therapy has a good prognosis but would require periodic plasma electrophoresis on follow up to pick up dissemination early.

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