Tumours of the odontoid peg revisited

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Abstract

Introduction: Tumours of the odontoid peg are rare but can potentially cause significant morbidity and mortality. Methods: A retrospective review of oncology and radiology database of tertiary orthopaedic oncology centre for all lesions affecting the odontoid peg over the last 12 years was performed. Results: We identified a total of 15 tumours involving the odontoid peg, majority being malignant. Myeloma was the most common tumour. Conclusion: Tumours of the odontoid peg are rare. Spinal surgeons and Radiologists need to be aware of them.

Key words: Odontoid; tumour; imaging

Introduction

The odontoid process (also known as dens) is a distinctive anatomical structure that arises perpendicularly from the upper surface of the axis (C2). It articulates with the atlas (C1), forming the pivot upon which the head rotates. The dens is related to many critical neurovascular structures, and pathologies involving the dens can lead to significant morbidity, and potentially mortality.[1] Pathological anomalies can be congenital, degenerative, inflammatory, traumatic or neoplastic in nature.

Tumours of the odontoid peg are rare and their diagnosis and management can pose a serious challenge to clinicians. We have reviewed our experience of odontoid peg tumours at our tertiary referral Centre for Orthopaedic oncology. We report the largest case series of tumours of the dens, and discuss the demographics and imaging features, to aid radiologists and orthopaedic spinal surgeons in the detection of such rare tumours.

Methods

A retrospective review of our oncology and radiology database for all lesions affecting the odontoid peg over the last 12 years was performed. Local committee approval was obtained as a service evaluation. All non-neoplastic lesions including fractures, inflammatory and degenerative lesions were excluded from our study. Primary diagnosis was made using MRI with or without CT. Definitive diagnosis was obtained following biopsies of odontoid lesions, performed within our tertiary radiology. In patients with metastasis, biopsy was performed of areas other than the odontoid, and in multiple myeloma cases, diagnosis was made in conjunction with a myeloma screening. All the images were subsequently reviewed by a musculoskeletal radiologist with 8 years’ experience.
Results

We identified a total of 15 tumours involving the odontoid peg. There was a mean age of 57 years (11–79 years) with a male to female predominance of 1.5:1 (9 males and 6 females). Four cases underwent definitive diagnoses with biopsies within our tertiary centre. Of the 15 cases, the majority (n = 12) were malignant and the remaining three cases were benign. The most common malignant tumor was multiple myeloma (six cases). Metastasis was second with a total of three cases. There were two chordomas, one case each of chondrosarcoma and ABC within our cohort, all of which were diagnosed definitively with biopsy within our department. The benign tumours in our cohort included two hemangiomas and an aneurysmal bone cyst (ABC) [Table 1].

Discussion

The odontoid process is an important structure that arises vertically cephalad from the body of C2 and attaches to the dorsal surface of the arch of the atlas; forming the atlanto-axial pivot joint.[2] It is held in place by three primary and one secondary ligament. The primary ligaments include the transverse ligament of the atlas (TL), which functions to limit anterior translation and flexion of the atlanto-axial joint, and two alar ligaments; situated ventral and cranial to the TL, that allow a 3–5 mm anterior shift of C1.[3] The main secondary ligament is the tectorial membrane (a continuous extension of the posterior longitudinal ligament of the vertebral column) that serves to limit over-flexion of the atlanto-axial joint. The odontoid process is related to many different anatomical structures including the spinal cord and vertebral artery. Any destruction of the dens can cause significant morbidity. Therefore, early diagnosis and management is essential.

Malignant

Multiple myeloma

Multiple myeloma (MM) is an idiopathic malignant disorder characterised by an abnormal proliferation of clonal plasma cells. It is the most common primary bone malignancy in adults, with a median age of diagnosis between 65 and 70 years.[4] Patients with suspected MM usually present with symptoms and signs typical of weight loss, fatigue (because of anaemia), renal insufficiency, hypercalcemia, recurrent infections and bone pain.[5,6] The spine is involved in approximately 60% of patients. This usually involves the thoracic region (between T4 and T6).[7] MM lesions of the dens are rare with very limited case reports in the literature.[8,9]

The diagnosis of MM depends on laboratory and imaging investigations. MM of the odontoid peg is evaluated best with MRI and CT. On T1, lesions typically appear as low signal lesion and on T2, the lesion is of high signal. On post-contrast sequences, myeloma deposits exhibit contrast enhancement in the dens.[10]

Radiographs and computed tomography (CT) play an essential role in the assessment of risk of pathological fractures and response to treatment, the latter being more sensitive. On both modalities, MMMs are sharply defined (or punched out) with endosteal scalloping [Figures 1 and 2].[11] In our case series of multiple myeloma of odontoid there were lucencies in the odontoid which consolidated with non-operative treatment (chemotherapy and brace).

The management of odontoid peg MM lesions depends on the degree of bony involvement and risk of fracture. Ideally, patients should be managed non-operatively with chemo-radiotherapy and high-dose steroids.[8,12] In some cases, Halo or occipitocervical fusions may be required to stabilise the spine.

Chondrosarcoma

Chondrosarcoma is the third most common malignant bone tumour arising from precursor cartilage forming cells, or secondary from a pre-existing benign osteochondromas and echondromas.[13,14] Chondrosarcomas account for 10% of all primary bone tumours, and have a 10% incidence in the spine.[15,16] They most commonly affect the thoracic spine, and clinical presentation is dependent on the tumour size, mass effect and involvement of adjacent neurovascular structures.[17,18] Chondrosarcomas of the odontoid peg, are very rare, and to the best of our knowledge, has not been reported in the literature. Elsewhere in the spine, almost all patients present with

Table 1: Tumor subtypes identified within our Cohort

<table>
<thead>
<tr>
<th>Tumour Subtype</th>
<th>N</th>
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<tbody>
<tr>
<td>Myeloma</td>
<td>6</td>
</tr>
<tr>
<td>Metastasis</td>
<td>3</td>
</tr>
<tr>
<td>Hemangioma</td>
<td>2</td>
</tr>
<tr>
<td>Chordoma</td>
<td>2</td>
</tr>
<tr>
<td>Chondrosarcoma</td>
<td>1</td>
</tr>
<tr>
<td>ABC</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>15</td>
</tr>
</tbody>
</table>

Figure 1 (A and B): Myeloma. CT sagittal (A) and coronal (B) pathological fracture through sclerotic myelomatous deposit (arrow)
pain and can have a palpable mass, with over 50% presenting with neurological symptoms.

The diagnosis of chondrosarcoma is highly dependent on correlation of radiological features with histological grading. On radiographs, chondrosarcomas appear as a destructive lesion with chondroid matrix. CT enables analysis of the adjacent structures, paravertebral extension, the pattern of calcification, depicted as the characteristic ‘rings and arcs’ appearance. MRI is useful in the assessment of extension of the tumour into the epidural and intraforaminal spaces. On T1WI, chondrosarcomas are of low signal and demonstrate lobulated high signal lesion on T2 with heterogeneous enhancement on post contrast images.

Chondrosarcomas respond poorly to chemo-radiotherapy, and surgical resection is the treatment of choice. The surgical management is beyond the scope of this study, however, it is worth mentioning that en bloc resection can be challenging in the management of chondrosarcomas of peg and therefore palliative decompression and radiotherapy may need to be considered.

**Chordoma**

Chordomas are extremely rare, slow growing malignant tumours, with an incidence of less than 0.0005%. They take origin from the notochord remnants, and most commonly arise in the clivus, spine and sacrocccygeal region. 50% of spinal chordomas are found within the cervical spine. Although they very rarely metastasize, chordomas are locally aggressive, often encapsulating the vertebral artery and cervical nerve roots. Due to the extensive nature of the disease and its slow progression, chordomas can be challenging to manage.

Like all other tumours mentioned on this paper, chordomas are diagnosed with a combination of radiological imaging and histopathological examination. CT and radiographic findings are nonspecific but may show a well-circumscribed calcified lesion with marginal sclerosis. On T1 chordomas may show intermediate to low grade intensity, with hyperintense signal on T2. Imaging features of chordoma and chondrosarcoma may appear similar, as both are hyperintense on T2. In one of our case of chordoma of odontoid, the tumour mass was causing cord compression. Biopsy is often required to clinch the diagnosis and planning of the biopsy route after consultation with spinal surgeon is crucial. The preferred approach for biopsy is lateral or posterolateral avoiding injury to vertebral artery.

Due to the rarity of these tumors, management is not standardized, but surgery was been widely accepted as the mainstay treatment of these lesions. Treatment options include en bloc resection or more commonly intracranial resection. Surgical management of odontoid peg lesions can be extremely challenging and may require a combined bilateral high anterior and posterior cervical approach. However, proton therapy is being commonly used for chordomas.

**Metastasis (Mets)**

The spine is the most common site of bony metastasis, with 10% found to affect the cervical vertebrae. The incidence of odontoid involvement is unknown. Spinal Mets can be osteolytic or osteosclerotic, depending on the primary lesion, of which 60% are caused by breast, prostate or lung cancer (Ca). These can be asymptomatic. If symptomatic, they can range from non-specific pain to symptoms of cord compression. Early detection is essential for the management and decrease morbidity.

Radiographic features are non-specific and can be lucent, sclerotic or mixed (lytic and sclerotic). MRI is the gold standard imaging modality for early detection and for assessment of the degree of cord compression. The signal of lesions depends on the nature of the primary disease (osteoblastic, lytic or sclerotic) and degree of mineralisation. Osteolytic lesions, such as lung or renal carcinoma show intermediate signals on T1WI and may be hyper or isointense on T2WI. Osteoblastic (prostate or thyroid Carcinoma) and mixed lesions (Breast Carcinoma or lymphomas) appear hypointense on T1 and T2WI. The metastasis in our series were from breast, renal and prostate carcinoma.
Treatment options for spinal metastasis are variable and depend on the patient’s symptoms, physiological status and spinal stability.\textsuperscript{[36]} Patients presenting with severe pain and stable spine may be treated with radiotherapy. Surgery, such as decompression or occipito-cervical fusion, is preserved for patients deemed fit for surgery, with significant spinal cord compression and instability.\textsuperscript{[36,37]} One patient in our series had occipitocervical fusion and the other two were managed with a halo.

**Hemangioma**

Hemangiomas are benign lesions of vascular origin formed by abnormal proliferation of capillary and venous vessels.\textsuperscript{[38]} They are the most common primary tumours of the spine,\textsuperscript{[39]} however, their incidence in the odontoid peg is unknown, and we could only identify one case report in the literature.\textsuperscript{[40]} Most hemangiomas are asymptomatic and often an incidental finding with an incidence as high as 27\% on MRI.\textsuperscript{[40,42]} 0.9–1.2\% of patients with hemangiomas are symptomatic with back pain and paraesthesia.\textsuperscript{[43]} Hemangiomas can rarely grow to considerable size, enough to cause a risk of pathological fractures. These can be associated with extraosseous component referred to as an
aggressive hemangioma. In these patients, imaging becomes critical, and is used in the evaluation of the site, size and degree of lysis.

On radiographs, hemangiomas affecting the vertebral bodies may appear as sclerotic lesions with a conspicuous trabecular pattern with vertical striations. CT imaging reveals the classical corduroy patterns consistent with hemangiomas and has the ability to evaluate the degree of bony involvement. MR imaging is the modality of choice in the assessment of soft tissue extension. Classic hemangiomas are hyperintense on T1 and T2 and low signal on STIR [Figure 6]. Atypical hemangiomas can be high on STIR and intermediate signal on T1 and T2. Chemical shift imaging can be helpful in diagnosing atypical hemangiomas with a signal drop off of over 20%. These can be associated with epidural component, the so-called aggressive hemangiomas. In our experience, atypical hemangiomas are often referred to our tertiary orthopaedic oncology centre as metastasis but by utilising chemical shift imaging to decipher intralsetional fat, a diagnosis of atypical hemangiomas can be made. This is especially important in odontoid as the alternative might be a percutaneous bone biopsy if it’s a solitary lesion. The treatment of hemangiomas is dependent on the size, site and symptoms. Patients who are at symptomatic may be offered endovascular embolization or percutaneous vertebroplasty, both of which have proven good results. Our patient with hemangioma of odontoid did not have any extrasosseus component and was managed symptomatically.

Aneurysmal Bone Cyst (ABC)
ABCs are benign aggressive tumours of unknown etiology, presenting 1.4% of all benign bony lesions. These are more common in the younger population, frequently in the first two decades of life and have a minor female predilection. The incidence of ABC of the spine is 3–30%. ABC of the odontoid peg specifically is even more rare with a limited number of cases reported in the literature. ABC of the odontoid peg presents with non-specific symptoms, including neck pain, neurological symptoms (secondary to cord compression). Therefore, imaging is required to delineate the diagnosis. On radiographs and CT, ABC appears as an expansile lucent lesion. There may be multiple internal septations, pathological fractures or collapse of the vertebral body. MRI is more specific and demonstrates a heterogeneous lesion with classical fluid–fluid levels [Figure 7]. However, fluid–fluid levels can be seen on other tumour subtypes, which include giant cell tumour, chondroblastoma. Treatment options of ABC of odontoid include embolization, sclerotherapy, cryo or radiotherapy and surgical resection with instrumentation [Figure 8].

Conclusion
Tumours of the odontoid peg are rare, with limited cases reported in the literature. Due to their scarcity, their diagnosis and management can be delayed. Spinal surgeons and Radiologists need to be aware of such lesions to initiate appropriate early diagnosis and management.

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Conflicts of interest
There are no conflicts of interest.
References


