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Thoracic mediastinal Chordoma an atypical location - Mimicking as Adenocarcinoma

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ABSTRACT

Background: Chordoma, a rare tumor accounting for less than 4% of primary bone tumors of which incidence of thoracic chordoma is 2-5%. Describing an incidental diagnosis of chordoma in a posterior mediastinal mass.

Case description: An asymptomatic 52year lady with an incidental posterior mediastinal mass was diagnosed Adenocarcinoma on endoscopic biopsy of the oesophagus. Nonresponsiveness to chemoradiotherapy warranted en-bloc resection with VATS- assisted thoracotomy which turned to be a Chordoma.

Conclusion: We report this atypical presentation of chordoma mimicking as adenocarcinoma with literature review — a unique learning experience.

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Introduction

Chordoma a rare malignant tumor of primitive notochordal origin accounts for 1-4% of all the bone malignancies[1,2]. It's a low-grade neoplasm which is highly recurrent, making it similar to malignant tumors[3,4]. Surveillance, Epidemiology, and End Results(SEER) database report an incidence of 0.08/100000 population with predominance in men by 2.7/1–1.6/1 in the age group of 50-70 year[5,6,7]. Chordomas mainly involve the axial skeleton with skull base (32%), sacrum (29.2%) and mobile spine (32.8%) of which thoracic chordoma form 2-5% [5,6,7].

We report a unique case of thoracic chordoma in a female mimicking poorly differentiated adenocarcinoma of esophagus.

Case report

A 52year old symptomatic lady referred to our center with an incidentally detected posterior mediastinal mass with no history of significance. She was diagnosed on a routine medical screening by esophagogastroduodenoscopy three years ago and was on yearly follow up. Increase in size noticed during the second year of follow-up on endoscopic ultrasonography. She was referred to Department of Gastrocolic surgery of our hospital, ultrasound done at our center showed 11.3 mm lesion of maximum extension, and the clinical diagnosis was Submucosal tumor. CT images showed 30X19mm sized ovoid, well-defined posterior mediastinum mass at D4-5 with bony erosion of the adjacent dorsal spine. MRI reconfirmed CT findings with intermediate signal intensities on both T1 and T2 weighted images. The mass was abutting the trachea and aortic arch without definite invasion and was indistinguishable from the esophagus (Fig 1). The differentials were Bronchogenic, Neurogenic, Submucosal tumor in order of probability.

Endoscopic biopsy of esophagus showed epitheliod tumor cells and mucin. Immunohistochemistry was positive for CK-7(Cytokeratin), P-63, Pan-cytokeratin, and faintly positive for CEA(Carcinoma embryonic antigen). These findings suggested of poorly differentiated adenocarcinoma.

Medical Oncology unit decided for adjuvant concurrent chemoradiation therapy (CCRT), and the patient received a total of 46 Gy radiation with Paclitaxel and Carboplatin. Follow-up PET-CT showed an increased standardized uptake value (SUV) at the lesion from 3.7 to 4.3 and also a hypermetabolic mass at D4 (Fig.2) based on which surgical management decided. The patient underwent VATS- assisted thoracotomy for mass excision and vertebral body en-bloc resection with a wide margin by the unit of Cardiothoracic and Spine unit of Orthopaedics (Fig 3).

Surgical specimen showed an ill-defined soft and gelatinous tumor (Fig 4A). Microscopically, epitheliod tumor cells arranged in cords or nets, and background was myxoid (Fig4B). Tumor cells had intracytoplasmic vacuoles with bubbly appearance, the so-called physaliferous cell (Fig4C). Immunostaining was positive for pan-cytokeratin and epithelial membrane antigen (Fig4D). Therefore, the tumor was diagnosed as Chordoma.

Given the changed diagnosis, after multi-department and patient consultation, it was agreed the management done was optimum and needed regular follow-up.

Discussion

The incidence of Chordoma spinal tumor at thoracic is rare and much rarer in the female. Few cases of Chordoma at the thoracic level reported in the literature[2,7-11] (Table1).

Radiological diagnosis based on the location was a gastric mucosal tumor, schwannoma, neurofibroma, ganglioneuroma, chondrosarcoma[2,11]. Our patient also had a slow and delayed growth in the lesion which is seen in conditions aforementioned[2,6]. Chordoma is similarly known to be a slow-growing, low-grade neoplasm which is locally aggressive, invasive, and radioresistant[6].

On exhaustive search of the literature, there is mention of epithelial neoplasms, metastatic adenocarcinoma known to mimic Chordoma histologically making diagnosis difficult[12-15]. One case report of the unlikely possibility of collision tumor of Chondroid chordoma and nasal adenocarcinoma in sino-nasal space by Gallet et al[16].

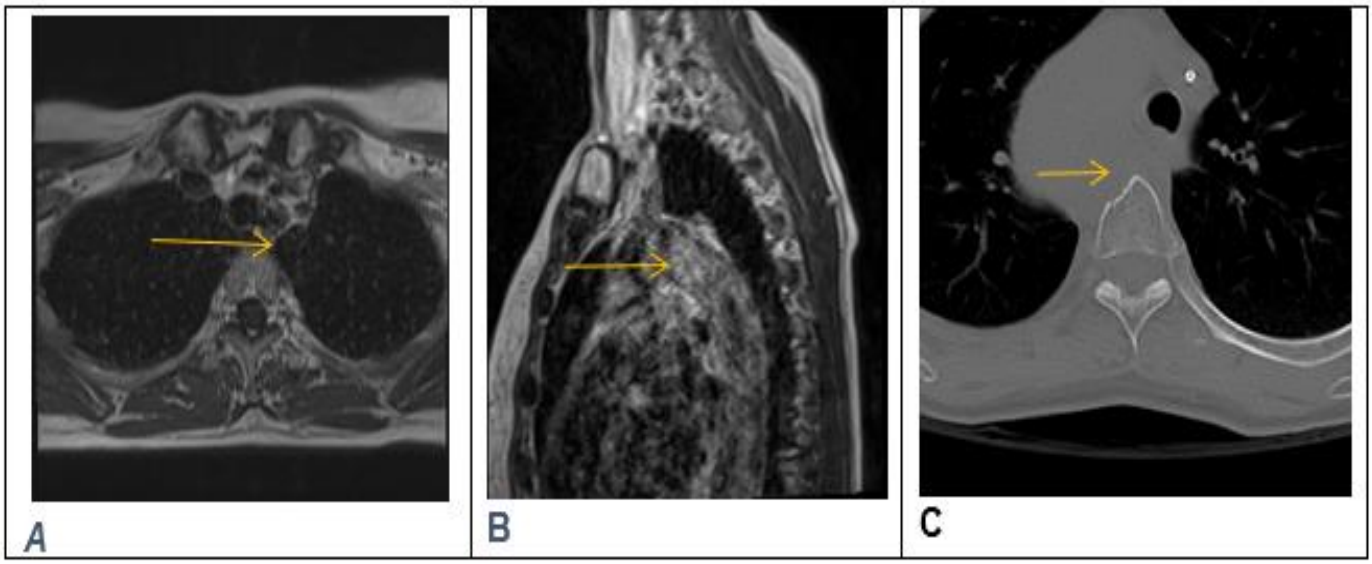

Fig.1 MRI and CT images of Dorsal spine reveal a circumscribed posterior medi-astinal mass (A) Axial T1-weighted image demonstrated that the mass lesion revealed intermediate signal intensity (B) Saggital image demonstrated mass indistinguishable from the esophagus (C) CT axial demonstrated bony erosion of the D4 body

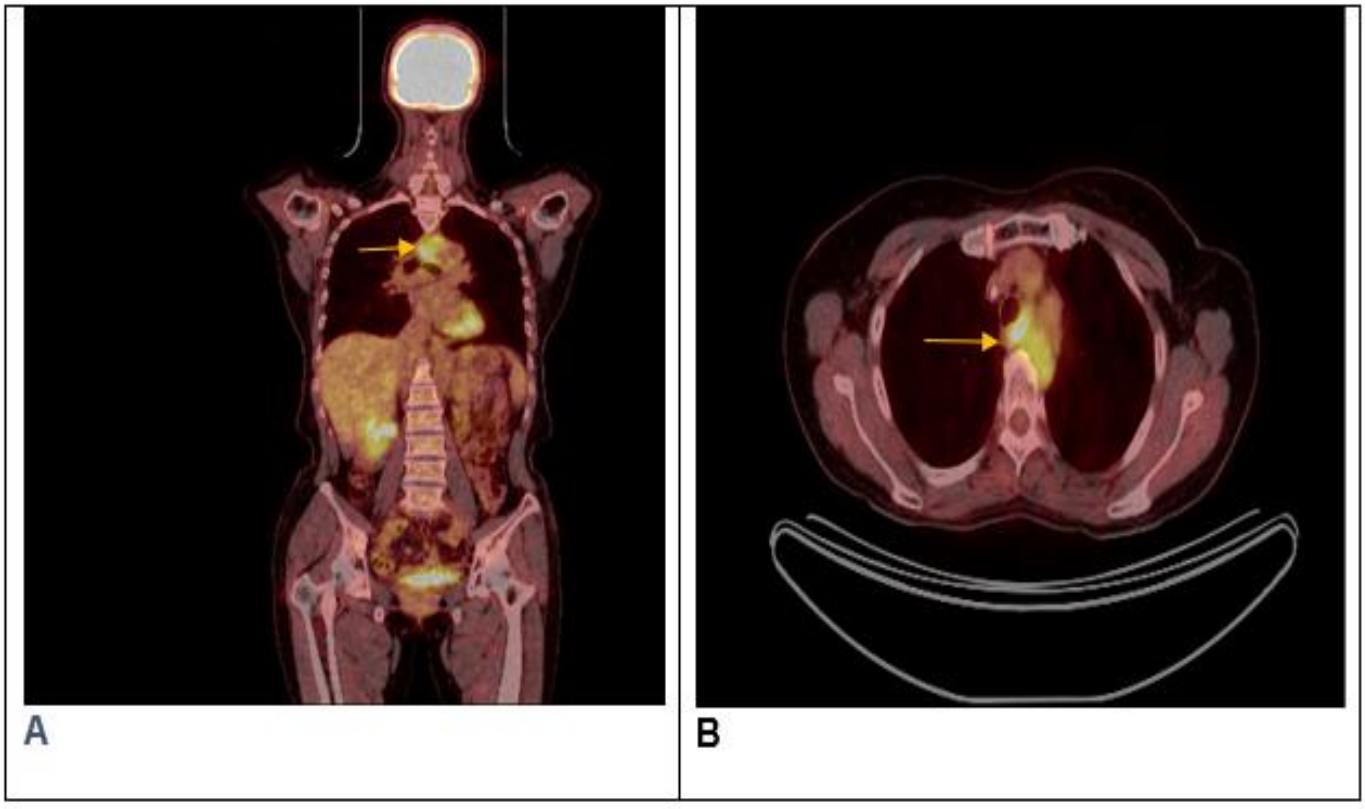

Fig 2. PET-CT showing increased SUV(Standardised uptake value) in Coronal (A) and Axial (B) images of the mass after CCRT.

Table1- Previous Case reports

Author	Year	Age	Sex	Complaints	Location
Walsh et al.	1992	69	F	Back pain, numbness	D10-12
Holden et al.	1998	20	M	Numbness, ataxia	D4-5
Hester et al.	1999	53	M	Dysphagia, dyspnea	C6-D1
Wang et al.	2008	25	F	Left hand anhydrosis	D1-5
Matsubayashi et al.	2012	47	F	Asymptomatic	Dorsal-Extraosseous
Royo et al.	2013	52	M	Dorsal pain	D5-7

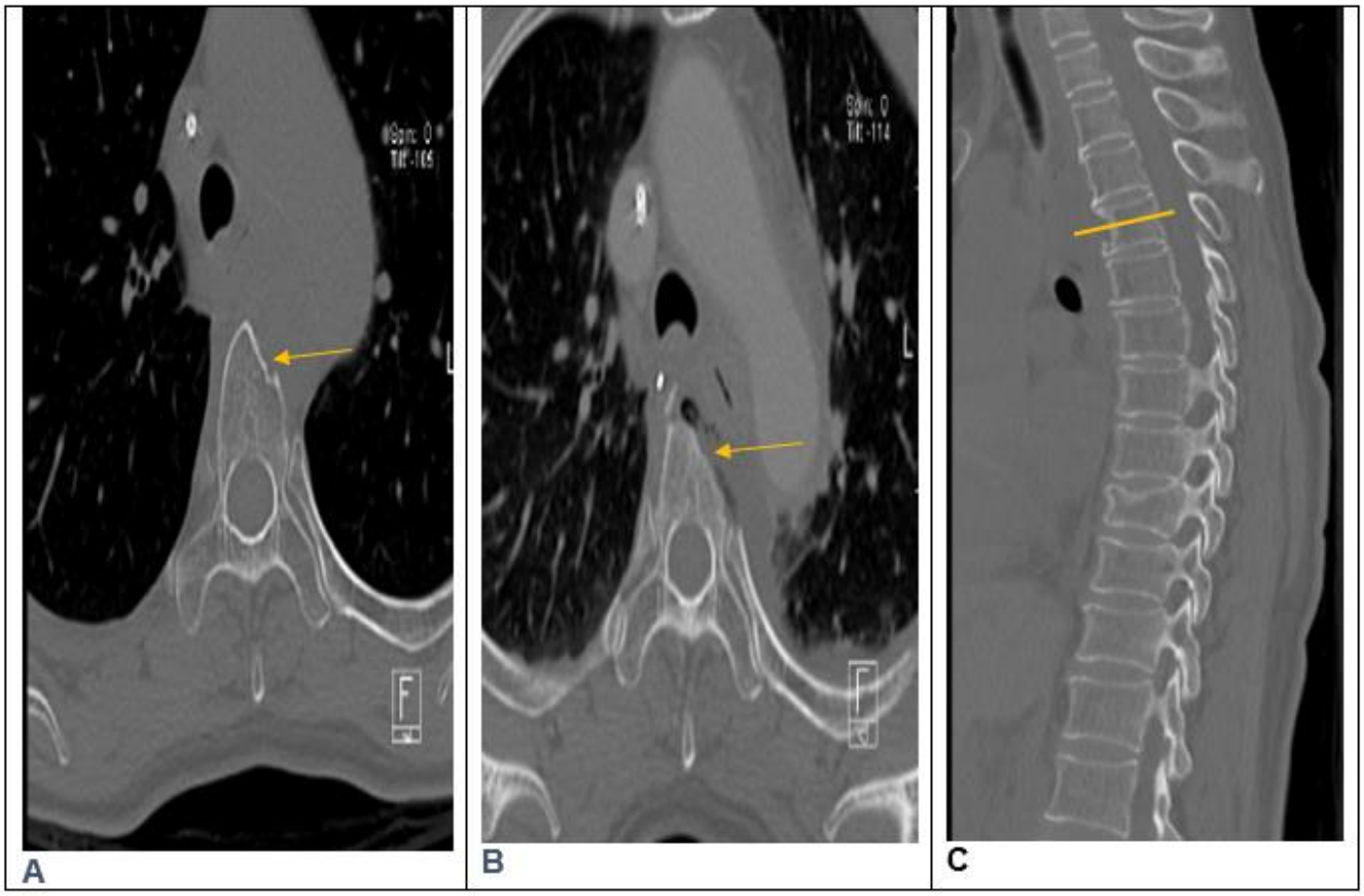

Fig 3. CT demonstrating (A) Axial image pre-operatively (B) Axial image post-operative en-bloc resection with a wide margin (C) Saggital scout image at the D4 level.

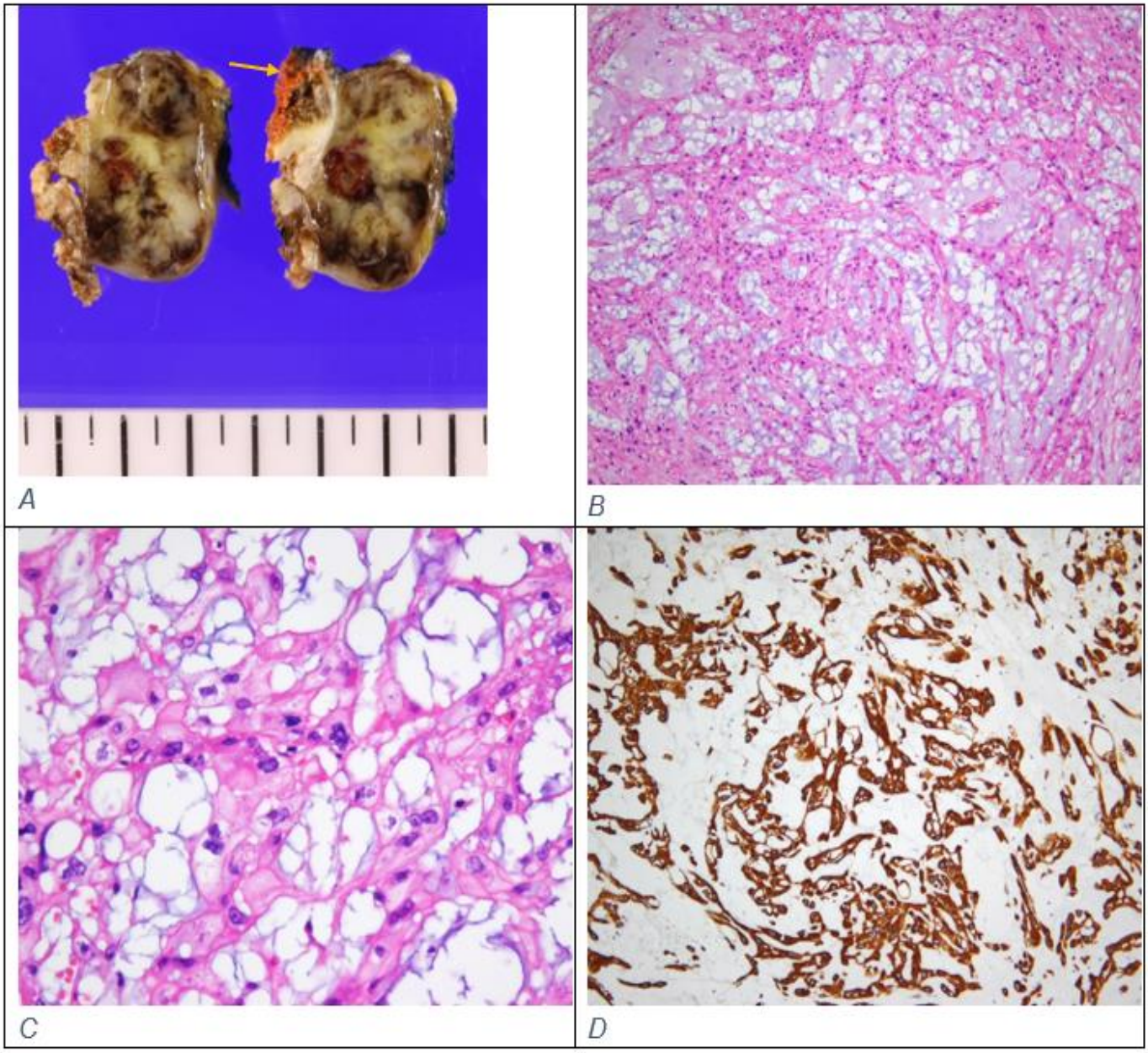

Fig 4. A. Gross photo of sagittal cut surfaces of extraosseous chordoma involving the anterior D4 vertebral bone. The tumor is soft, gelatinous, tan-gray, and hemorrhagic. B. The tumor shows epithelioid tumor cells arranged in cohe-sive cords and nests. The stroma is basophilic and myxoid (x100). C. Tumor cells have abundant eosinophilic cytoplasm and intracytoplasmic vacuoles (arrow) with bubbly appearance (x400). D. Tumor cells are diffusely positive for pancytokeratin(x100).

Histologically chordoma is classified into Classic, chondroid and undifferentiated. The classic chondroma is typical of the extensively abundant vacuolated cytoplasm known as Physaliphorous cell[6].The cytokeratin staining has sensitivity and specificity of 98%,100% for chordoma differentiating from chordosarcoma[6,17]. In retrospect when the histopathological slides of FNA reanalyzed traces of Chordoma features were present but missed due to small biopsy sample from the site.

True to its nature, the tumor was radioresistant to CCRT and noticed to be locally aggressive on PET-CT even after a high dose of radiation of 48 Gy[6,18-20]. The lesion was excised en-bloc which is the treatment of choice for Chordoma[2,6,21].

Conclusion

Posterior mediastinal Chordoma at the thoracic level is a very rare. Owing to its immune-histological mimicking is tricky to diagnose and can be missed. Management is a challenge due to its anatomic location and added local invasion for which a multidisciplinary approach is needed, taking the patient into confidence to provide the optimum treatment for the best possible outcome.

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