A Case of Metastatic Dedifferentiated Chordoma  
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**INTRODUCTION**
- Chordomas are rare, locally aggressive bone tumors.  
- They arise from the embryonic notochord.  
- Occur frequently in the skull base or sacrum.  
- Incidence: 0.1/100,000/year, common in men.  
- Subtypes: Classical, Chondroid and Dedifferentiated.  
- Dedifferentiated type has the worst prognosis.

**CASE PRESENTATION**
- 20 year old female with no significant past medical history presented with 3 months of worsening back pain.  
- MRI C-spine showed diffuse bone marrow signal.  
- CT abdomen and pelvis showed lytic lesions throughout the axial skeleton and hepatic lesions.  
- CT chest showed multiple pulmonary nodules and right cervical lymphadenopathy.  
- She underwent bone marrow biopsy and excisional biopsy of the right neck mass.  
- Right neck mass: tumor cells are positive for brachyury by IHC, feature of dedifferentiated chordomas.

**IMAGING**

**PATHOLOGY**

**DISCUSSION**
- Unresectable chordomas are treated with systemic therapy.  
- A prospective phase II study demonstrated modest responses with Imatinib, a tyrosine kinase inhibitor as advanced chordomas express PDGFR $\beta$ (Stacchiotti et al., 2012).  
- Anthracyclines, Alkylating agents, Platinum agents, Topoisomerase II inhibitors, Vinca alkaloids were experimented in few case reports with moderate success (Meis, Raymond, Evans, Charles, & Giraldo, 1987)(Fleming et al., 1993)(Chou et al., 2009).

**CLINICAL COURSE**
- Foundation one testing with no actionable mutations.  
- She was started on Imatinib but progressed.  
- Received 3 cycles of Doxorubicin and Ifosfomide.  
- Presented with spontaneous bleeding into liver lesions with disease progression.  
- Hospital course complicated with hemoperitoneum and DIC.

**CONCLUSIONS**
- This case illustrates the aggressive nature of dedifferentiated chordomas.  
- There is no general consensus on treatment given their rarity.  
- More research is needed to identify novel targeted therapies as conventional treatments offer a limited benefit.