

A Case of Metastatic Dedifferentiated Chordoma

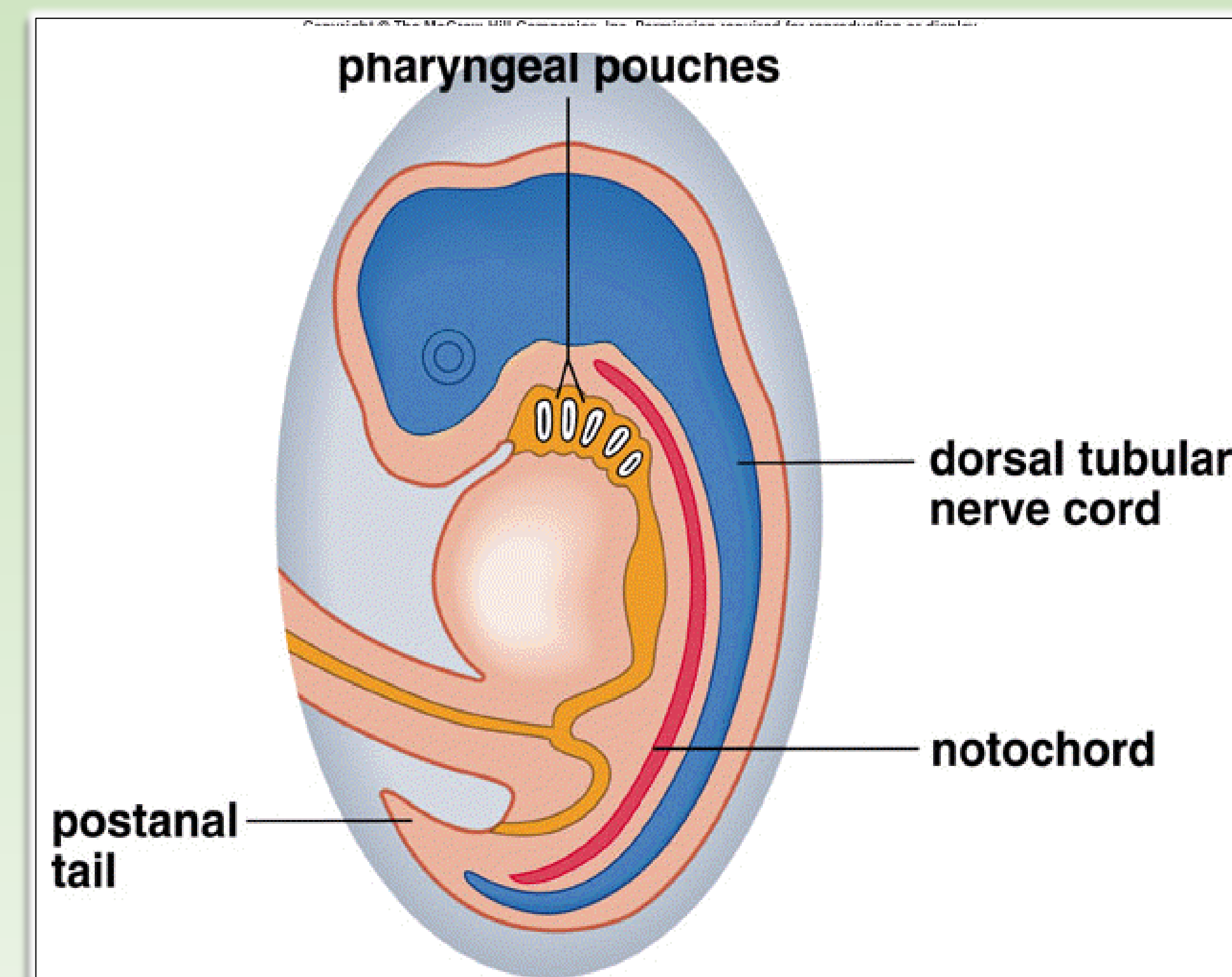
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INTRODUCTION

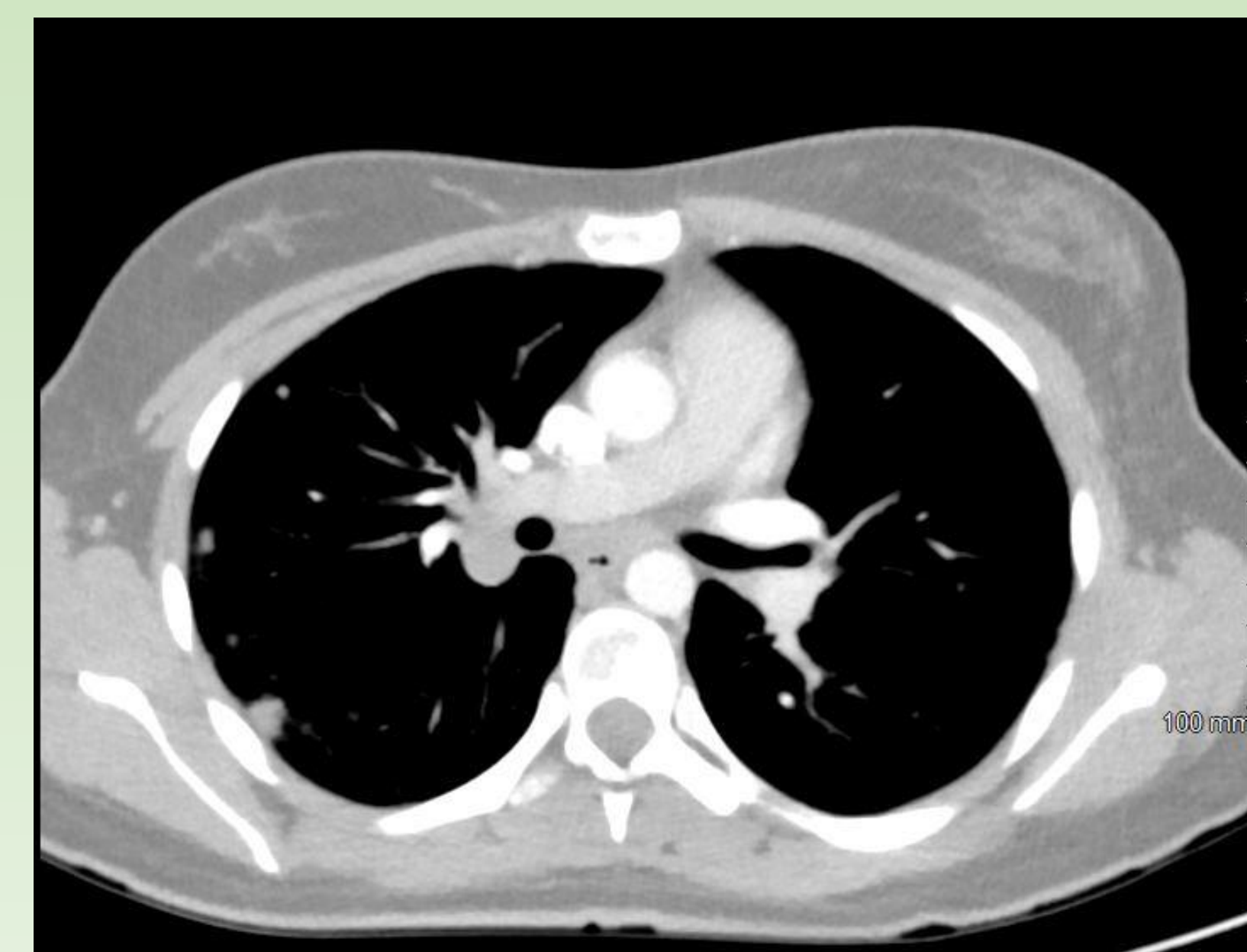
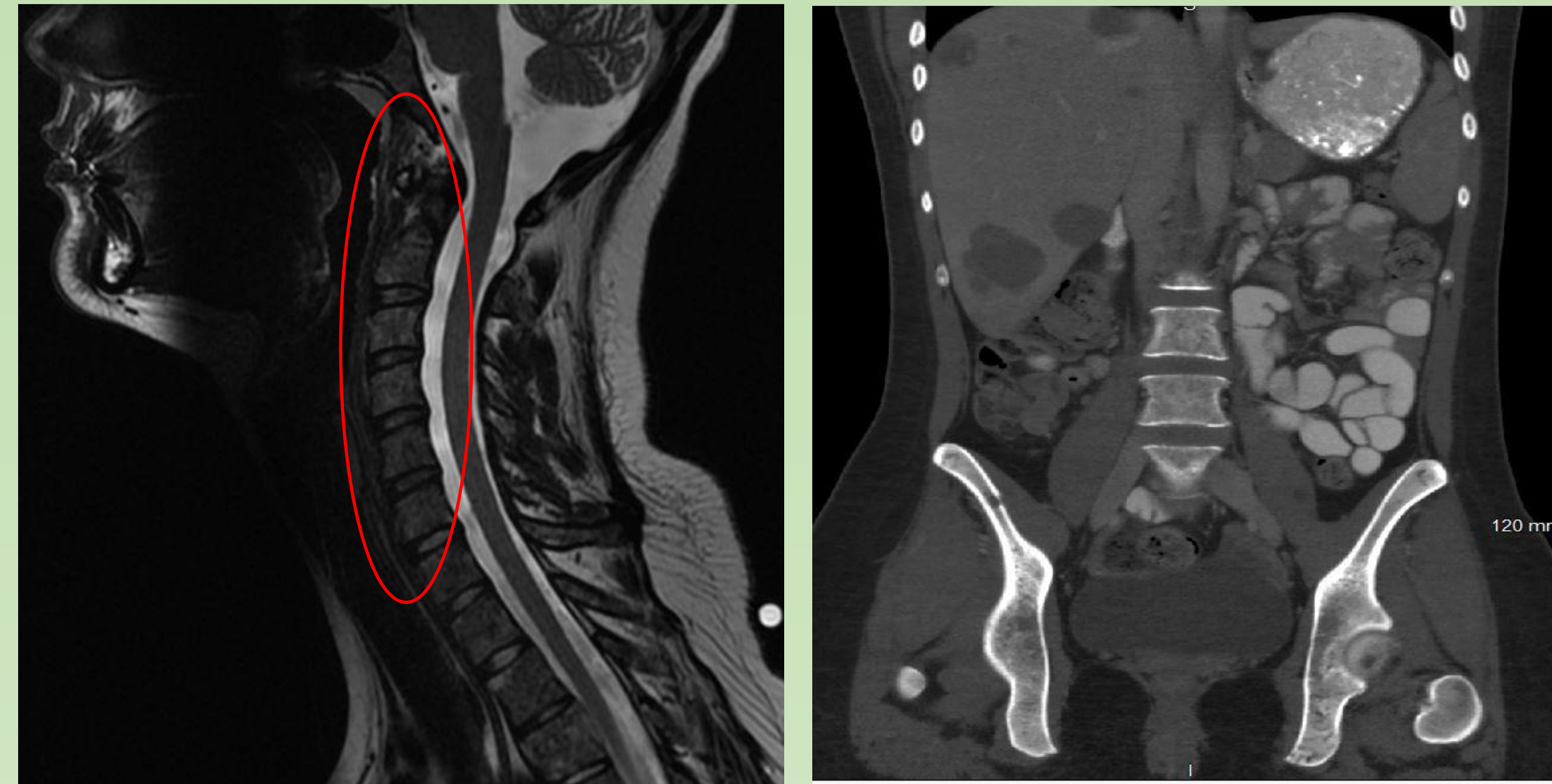
- Chordomas are rare, locally aggressive bone tumors.
- They arise from the embryonic notochord.
- Occur frequently in the skull base or sacrum.
- Incidence: 0.1/100,000/year, common in men.
- Subtypes: Classical, Chondroid and Dedifferentiated.
- Dedifferentiated type has the worst prognosis.



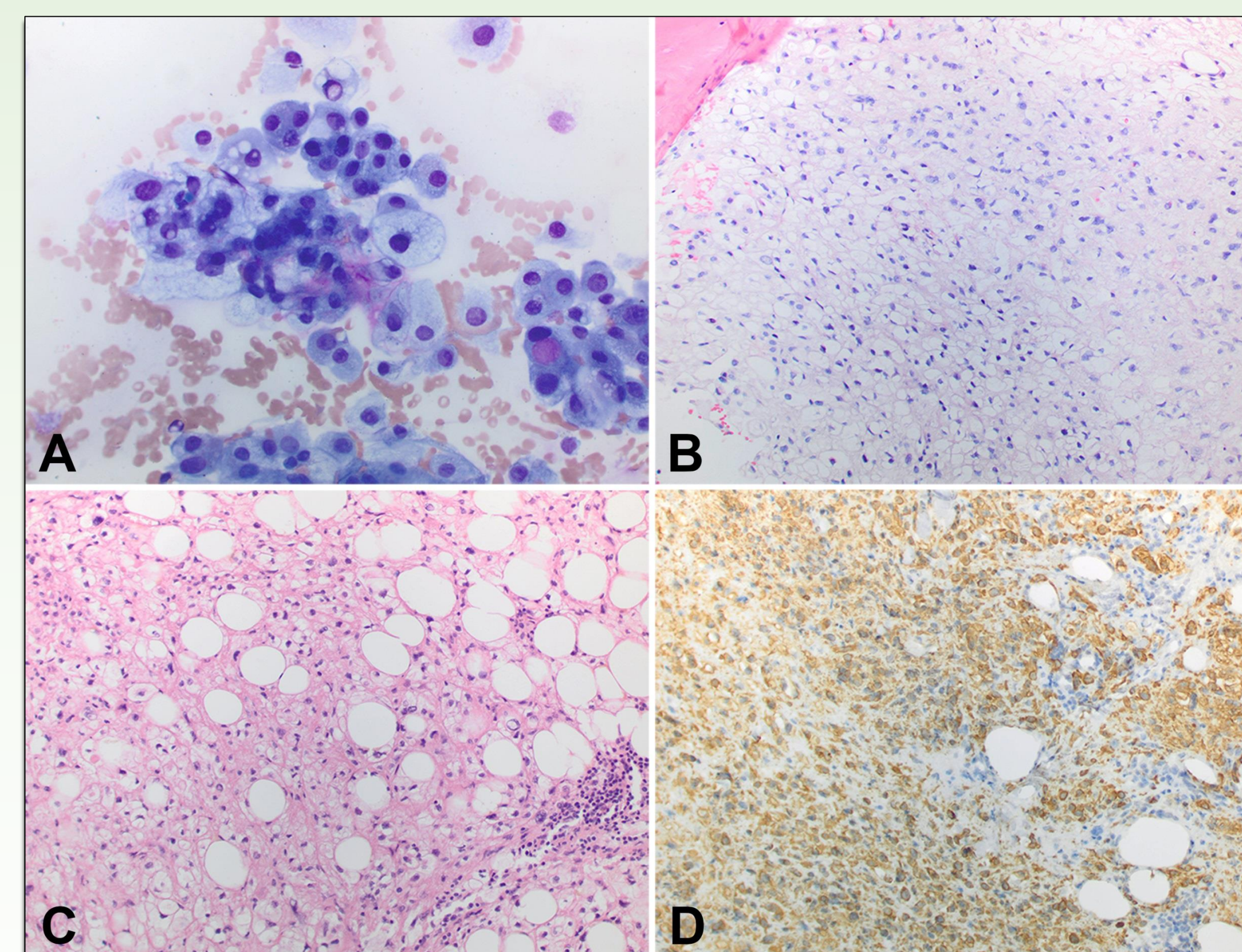
CASE PRESENTATION

- 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



- Bone marrow aspirate smear showing numerous epithelial cells with vacuolated cytoplasm.
- Complete replacement of the bone marrow with vacuolated epithelial cells.
- Soft tissue neck mass with vacuolated epithelial cells.
- Soft tissue neck mass stained positive for keratin.

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.

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 β (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).
- Patient was started on combination chemotherapy with Doxorubicin and Ifosfomide after disease progression on Imatinib.
- Unfortunately, she had an aggressive clinical course.

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.