



INTRODUCTION

Fibrous dysplasia is a rare, chronic benign bone disorder in which normal bone is replaced by abnormal fibrous tissue, resulting in structurally weakened bones and an increased risk of deformities and fractures. It is caused by a gene mutation on chromosome 20, affecting the region of Gs- α subunit, and leading to an overproduction of cyclic adenosine monophosphate (cAMP). This molecular alteration disrupts osteoblastic differentiation resulting in disorganized fibro-osseous tissue formation. Radiologically and histologically, fibrous dysplasia may mimic other benign and malignant bone lesions, including ossifying fibroma, osteblastoma, and low-grade osteosarcoma, fibrosarcoma, and chondrosarcoma. Although malignant transformation of fibrous dysplasia is rare, it must be considered. Hence, distinguishing fibrous dysplasia from these entities is critical, as early malignant lesions may lack overtly aggressive features. Careful evaluation for malignant osteoid or cartilage production, increased cellularity, cytologic atypia, and infiltrative growth is essential to ensure accurate diagnosis and appropriate clinical management.

ROLE OF THE PATHOLOGISTS' ASSISTANT

Following surgical resection of the left eighth rib, the specimen was received in the pathology laboratory. The Pathologists' Assistant (PA) oriented, measured, and radiographed the specimen, inked the margins, and systematically gross examined the rib, with careful assessment of cortical integrity and possible soft tissue extension.

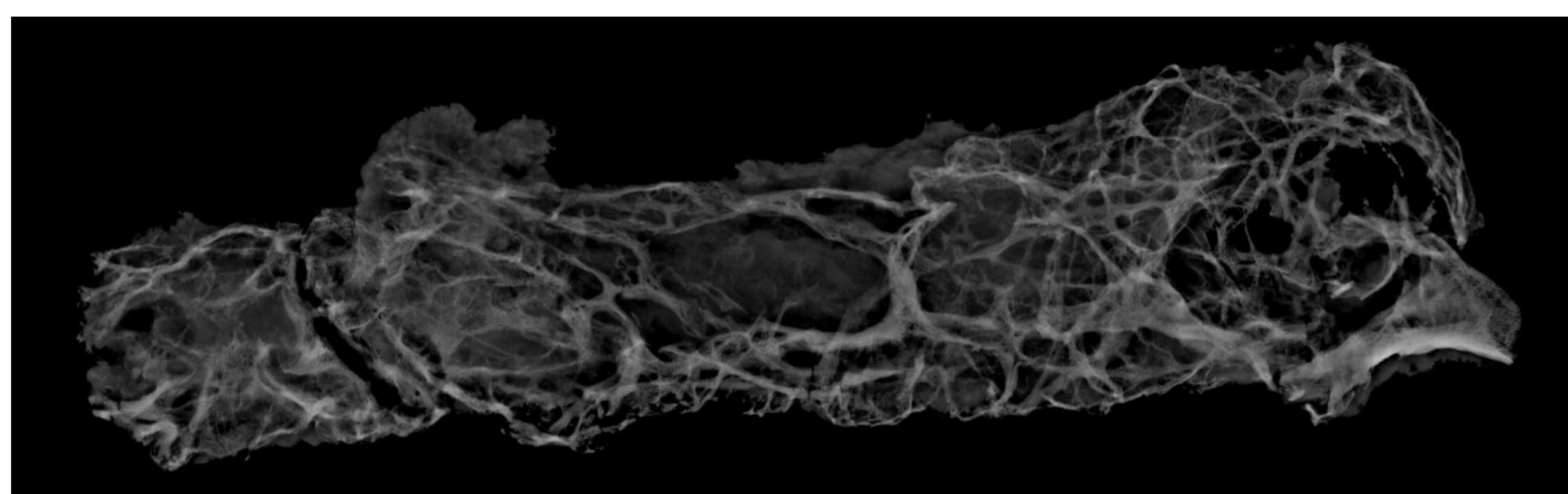


Figure 1. Radiograph of the rib segment demonstrating a lucent, lobulated and expansile lesion with multifocal areas of cortical penetration.

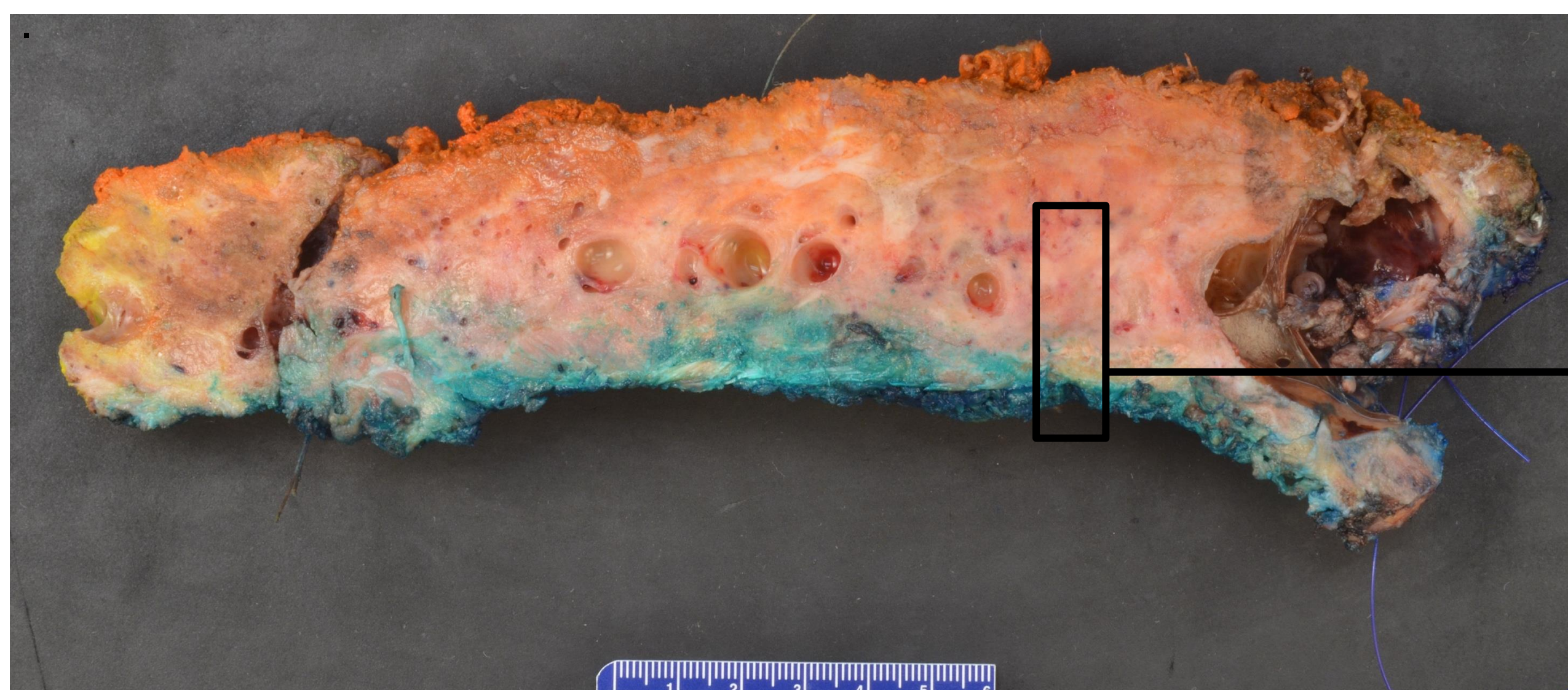


Figure 2. Cut surface of rib segment showing different components of the lesion and its relation to margins and pleura. Legend: orange - superior, green - inferior, yellow - anterior, blue - posterior

Gross description: The rib was diffusely distorted with loss of normal bony architecture. The lesion was predominantly calcified with scattered softer areas and demonstrated fibrous and cystic changes measuring 0.2 to 5.0 cm, extending beyond the cortex into surrounding soft tissue and exerting mass effect on the pleura. The lesion appeared to abut the superior, anterior, and lateral margins, was 0.3 cm from the inferior margin, and 0.5 cm from the posterior margin. A full-thickness sutured surgical defect was noted 5.0 cm from the anterior margin, and the pleura appeared smooth without evidence of invasion.

Representative sections were submitted from all margins and areas of interest. The anterior and posterior margins were submitted en face. The superior margin, inferior margin, and the lateral margin were submitted perpendicularly. Sections of the pleura and the fibrous and cystic areas were extensively sampled. All tissue was processed routinely, decalcified as required, embedded in paraffin, sectioned, and stained with hematoxylin and eosin for microscopic evaluation.

PATIENT HISTORY

- 52 year old male patient, history of thyroid cancer, thyroidectomy + lymph node dissection.
- In 2013, the patient was found to have a posterior left eighth rib monostotic lesion that was biopsied and initially interpreted as bone dysplasia, with imaging demonstrating stability dating back to 2008 and thus managed with consistent observation.
- Over the subsequent decade, the patient experienced progressive rib discomfort and weight loss (60 lb).
- Computed tomography (CT) scans demonstrated enlargement of the lesion involving the entire left eighth rib, associated with expansile bone destruction, rib displacement, and suspected pathologic fracture.
- A second biopsy was performed and was consistent with fibrous dysplasia, however, given the extent of the lesion, it was difficult to rule out a more aggressive lesion.
- Considering the patient's young age, symptomology, and the lesion progression, surgical resection was performed.

RESULTS

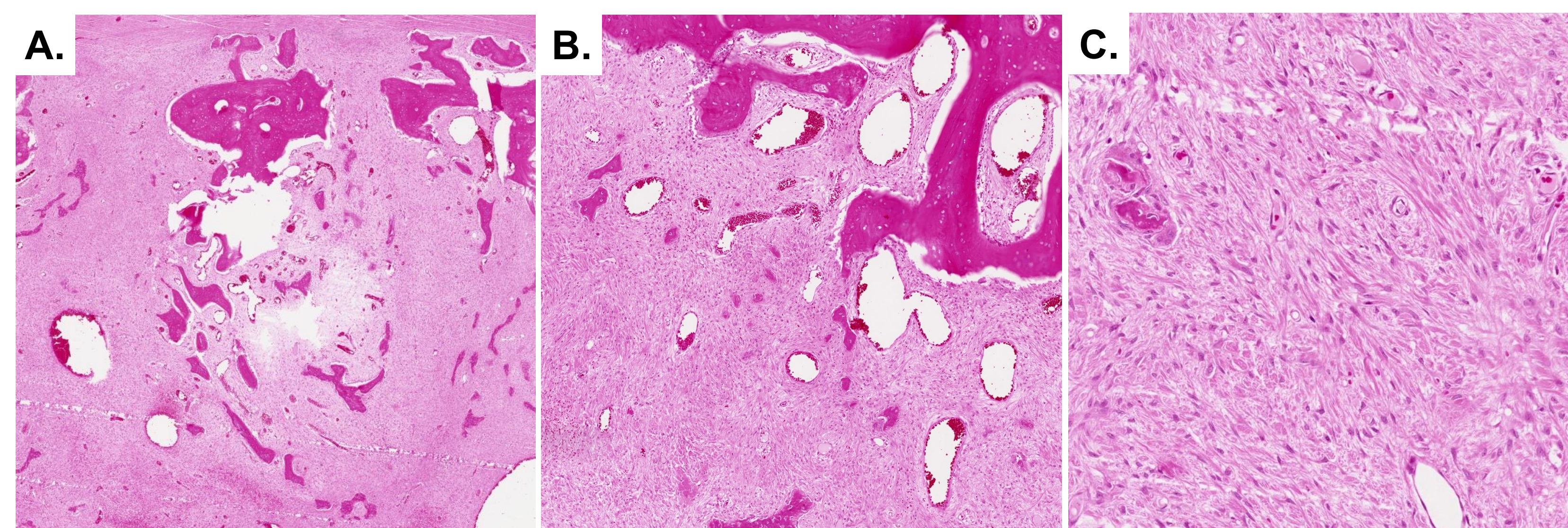


Figure 3. H&E-stained perpendicular section demonstrating the fibro-osseous lesion with cortical involvement at increasing magnification: (A) 4x, (B) 10x, and (C) 20x.

Microscopic examination demonstrated a fibro-osseous lesion involving the entire extent of the rib segment with both solid and cystic components. The solid areas had variable cellularity and contained scattered irregular woven bone without osteoblast rimming, surrounded by sheets of bland spindle cells within a vascular stroma. No necrosis or significant mitotic activity was identified. Foci of hemorrhage and hemosiderin deposition were present, and some areas showed extension into the cortex and adjacent soft tissue. The cystic spaces lack a defined epithelial lining. Overall, the morphologic features are consistent with fibrous dysplasia, with no evidence of malignant transformation, metastatic disease, or another primary bone or hematopoietic neoplasm. However, given the clinical history, progressive growth over approximately 12 years, and soft tissue extension, the case was referred to the Bone and Soft Tissue service at Mount Sinai Hospital for expert consultation, with findings to be reported in a supplemental report.

CONCLUSION

This case illustrates an atypical clinical course with interval enlargement over more than a decade, highlighting the importance of thorough gross examination and extensive sampling to exclude early malignant transformation. For Pathologists' Assistants, meticulous orientation, margin assessment, and representative sampling of areas demonstrating cystic change, cortical breach, or soft tissue extension are critical to ensure accurate histologic evaluation. Although the grossing approach in this case allowed comprehensive assessment of margins and lesion heterogeneity, similar cases may benefit from radiologic-pathologic correlation and targeted sampling of radiographically suspicious regions. Continued documentation of long-standing or atypically progressive fibrous dysplasia cases may further refine diagnostic criteria and inform best practices in specimen handling and sampling for fibro-osseous lesions.

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