

*Orthopaedic • Radiology • Pathology Conference*

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**Elbow Mass in a 9-year-old Girl**

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**HISTORY AND PHYSICAL EXAMINATION**

A 9-year-old girl presented with a history of swelling in her right elbow. There was a gradual loss of exten-

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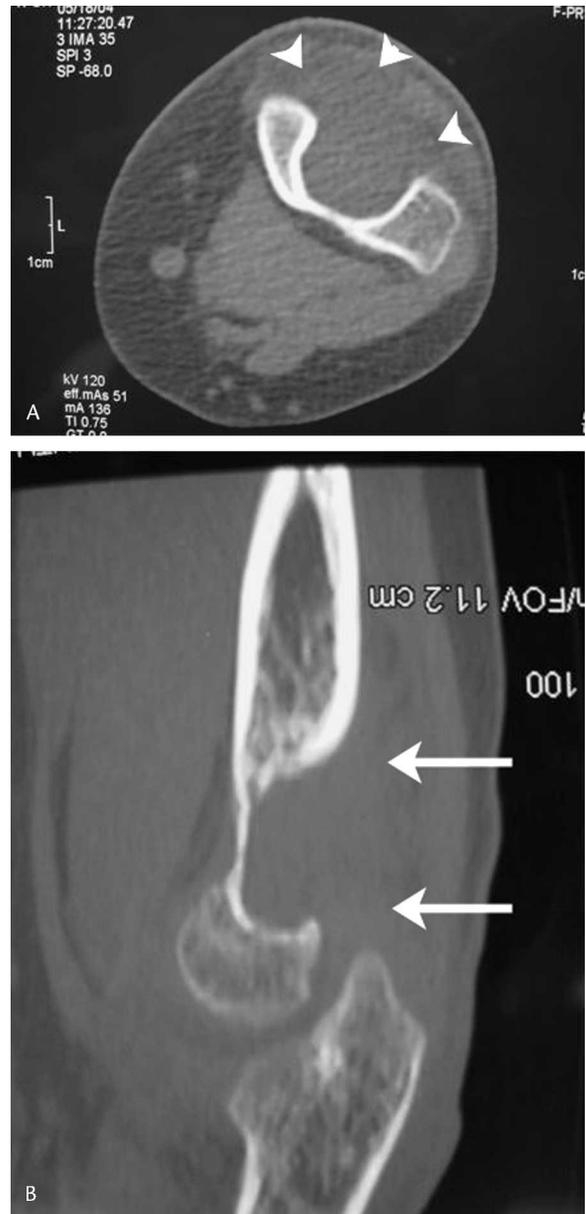
sion over a period of 6 months. The patient had minimal discomfort. There was no history of fevers or trauma to the area. Physical examination demonstrated soft swelling along the lateral and posterolateral aspect of the elbow in the region of the radiocapitellar joint. The child lacked 20° of active and passive extension but had full flexion. Motion of the elbow was painless. There was no change in the size of the soft tissues with position of the extremity.

Plain films (Fig 1), computed tomography (CT) (Fig 2), and magnetic resonance imaging (MRI) (Fig 3) were performed.

Based on the history, physical examination, and imaging studies, what is the differential diagnosis?



**Fig 1A–B.** Plain radiographs of the right elbow demonstrate a prominent and enlarged olecranon fossa (A) on the anteroposterior view (B). No obvious effusion was noted.



**Fig 2A–B.** (A) Axial CT image demonstrates a mass with soft tissue attenuation (arrowheads) noted extending posteriorly from the olecranon fossa. (B) On the sagittally reconstructed CT image, the mass (arrows) appears to be remodeling and deepening the olecranon fossa. There is no evidence of periosteal reaction or bony destruction.



**Fig 3A–B.** (A) Coronal and (B) sagittal 0.3-T open-gap MR images without intravenous contrast of the elbow demonstrate a heterogeneous hyperintense mass lesion (arrows) on T2\*-weighted gradient echo images involving the olecranon fossa resulting in remodeling of the adjacent osseous structures.

## IMAGING INTERPRETATION

Plain radiographs of the right elbow demonstrated a prominent and enlarged olecranon fossa on the anteroposterior view (Fig 1A) with suggestion of soft tissue fullness posteriorly in the lateral projection (Fig 1B). No obvious effusion was noted.

On CT, a mass with soft tissue attenuation was noted extending posteriorly from the olecranon fossa (Fig 2A). The mass appeared to be remodeling and eroding the fossa (Fig 2B). There was no evidence of periosteal reaction or bony destruction, and there were no calcifications in the mass.

A whole-body bone scan (images not available) demonstrated solitary focal uptake of radiopharmaceutical within the right elbow.

MRI demonstrated the mass measured  $4.7 \times 3.1 \times 2.1$  cm in maximal dimensions and was predominantly hetero-

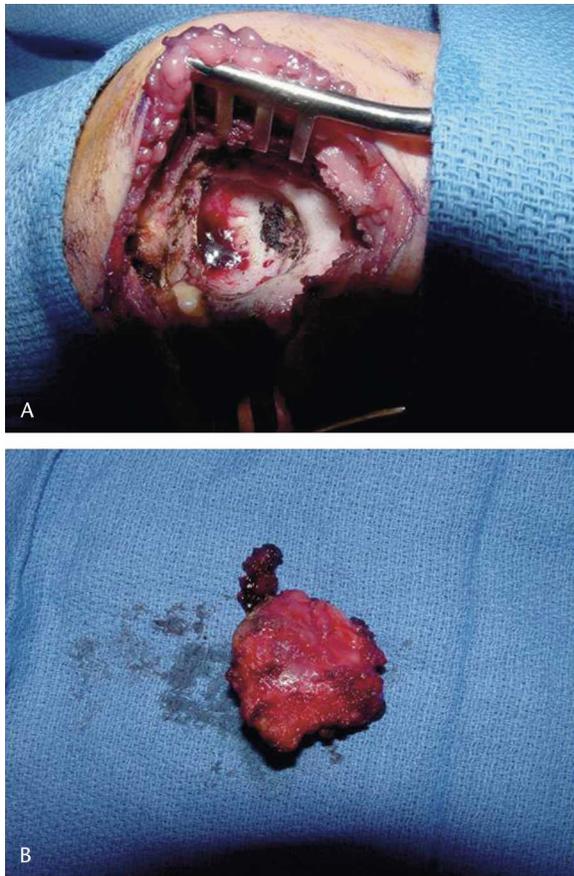
geneously hyperintense on T2\*-weighted gradient echo images. We noted no substantial reactive edema in the marrow or adjacent soft tissues (Fig 3).

## DIFFERENTIAL DIAGNOSIS

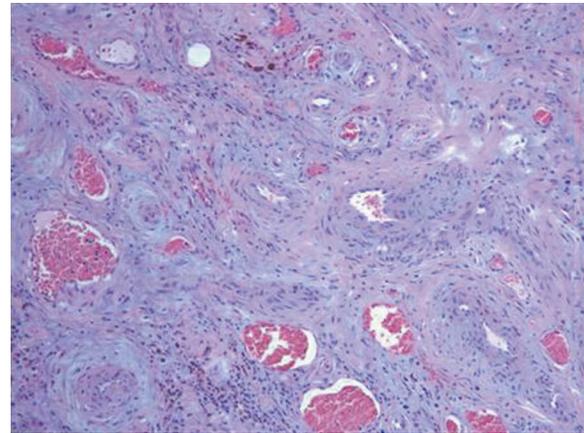
- Pigmented villonodular synovitis
- Synovial chondromatosis
- Inflammatory arthritis with pannus formation
- Chronic infection of mycobacterial origin
- Synovial sarcoma
- Synovial hemangioma

A CT-guided biopsy was performed, the soft tissue mass was then excised, and the histology of the mass was examined (Figs 4, 5).

Based on the history, physical examination, imaging studies, and histology, what is the diagnosis and how should this lesion be treated?



**Fig 4A–B.** Intraoperative photographs demonstrate (A) the marked remodeling and enlargement of the olecranon fossa and (B) the gross sample of the removed synovial hemangioma with focal regions of bleeding and hemosiderin.



**Fig 5.** A photomicrograph demonstrates variable-sized thin-walled vascular channels (cavernous type), thick-walled veins, and capillaries within intervening fibrous septae. Focal hemosiderin deposits are also noted (Stain, hematoxylin and eosin; original magnification,  $\times 100$ ).

*See page 241 for diagnosis and treatment.*

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## HISTOLOGY INTERPRETATION

Samples from the CT-guided biopsy revealed synovial-lined tissue with small-caliber blood vessels. Also seen were thick-walled large vessels suggestive of a benign vascular lesion, such as a hemangioma or an arteriovenous malformation.

Subsequently, the patient underwent resection of the soft tissue mass (Fig 4A). The gross specimen was a somewhat round, nodular, tan-brown soft tissue mass measuring  $3.5 \times 3.5 \times 2.0$  cm, which on cut surface revealed areas of pinpoint bleeding suggestive of blood vessels (Fig 4B).

The microscopic examination revealed a benign vascular lesion of the subsynovial lining composed of capillary-sized vessels arranged in large groups and separated by thin connective tissue septa. Small foci also demonstrated cavernous-type dilated vessels and thick-walled veins. Focal hemosiderin and small fatty areas were also present within the hemangiomatous tissue (Fig 5).

## DIAGNOSIS

Synovial hemangioma, mixed capillary and cavernous type

## DISCUSSION AND TREATMENT

Synovial hemangiomas are very uncommon, benign lesions, representing less than 1% of all hemangiomas. Approximately 60% of synovial hemangiomas are found in the knee; however, they can occur in almost any joint.<sup>3,11-14,16,18,21,23,27</sup> They are predominately found in children and young adults and commonly manifest as swelling and joint pain.<sup>15,24,26</sup> They are also a cause of spontaneous hemarthrosis and monoarthritis in the pediatric and adolescent population. Posterior interosseous nerve palsy secondary to compression from a synovial hemangioma has also been described.<sup>5</sup> Depending on the size of the hemangioma, a tender, spongy or firm mass may be palpable. Upon elevation of the extremity, the mass may be noted to decrease in size, related to venous emptying. Although an uncommon entity, early diagnosis of synovial hemangioma is important because recurrent hemarthrosis may lead to chronic inflammatory synovitis and joint damage.<sup>1,6</sup>

The appearance of the lesion in this case was consistent with a somewhat indolent intraarticular process likely of synovial origin. Differential diagnoses included pigmented villonodular synovitis; synovial chondromatosis; inflammatory pannus secondary to arthritides, such as juvenile onset rheumatoid arthritis; chronic mycobacterial infec-

tion; and synovial sarcoma. Certain clinical and imaging features are useful in limiting the differential diagnosis.

Pigmented villonodular synovitis is a slow-growing benign proliferation of the synovium of uncertain etiology. It typically occurs in the second to fourth decades of life. Plain radiographs and CT may demonstrate the presence of a high-density effusion and erosions, particularly in the lower volume joints. On MRI, there is typically some low signal on all sequences with a phenomenon known as "blooming" on gradient echo sequences due to the presence of hemosiderin. Though there was erosion in our case, the lack of concomitant effusion and low signal on MRI, as well as the atypically young age of the patient, made this diagnosis less likely.

Synovial chondromatosis is a benign condition marked by metaplastic proliferation of the synovium. Similarly to pigmented villonodular synovitis, it can result in erosions in the lower volume joints. Purely cartilaginous bodies are well delineated on MRI or MR arthrogram and tend to be high signal on T2-weighted images. The bodies may become calcified or ossified (synovial osteochondromatosis), making the diagnosis possible on plain radiographs or CT scan. It usually occurs in the third to fifth decade of life and is 2 to 4 times more common in male patients.<sup>7,23,25</sup> These clinical features make it less likely in our case.

Both chronic mycobacterial infection and pannus associated with inflammatory arthritis may present as intraarticular masses not unlike that in this case. However, juvenile rheumatoid arthritis is usually polyarticular in nature and both entities are characterized by loss of joint space, reactive joint effusion, articular erosions, and marrow edema, which were not seen in this case. Although juvenile rheumatoid arthritis has been detected in patients as young as 5 year old, most patients with juvenile rheumatoid arthritis present after age 10. Mycobacterial infection in an otherwise healthy child from a nonendemic area would be unlikely.

Synovial sarcomas can rarely be intraarticular in location (< 10%). However, they are associated with calcification in 1/3 of cases and are typically seen in patients 15 to 35 years of age.

Conventional radiographs are normal in approximately 1/2 of the cases of synovial hemangiomas.<sup>10</sup> Radiographs may demonstrate a nonspecific soft tissue mass. Localized phleboliths may also be seen but are rare. In our case, plain radiographs were actually beneficial and demonstrated marked scalloping and remodeling of the olecranon fossa of the humerus. Other associated findings, such as osteoporosis, periosteal reaction, and early epiphyseal maturation, secondary to the hyperemia, have been described but are uncommon.<sup>10</sup> Numerous episodes of intraarticular hemorrhage may simulate hemophilic arthropathy.

CT examination may also demonstrate a soft tissue mass and small calcifications or phleboliths. In our case, CT accurately depicted the chronic remodeling of the distal humerus; however, no phleboliths were evident. Sonographic examination of the affected joint may demonstrate a vague mass.

The preferred modality for assessing these lesions is MRI and their appearance is often fairly characteristic.<sup>2,17,19,22</sup> T1-weighted images typically demonstrate a poorly marginated mass that is isointense to skeletal muscle. Increased T1 signal may be present within the mass, corresponding to fat signal. T2-weighted images demonstrate a mass that is markedly hyperintense in signal, likely secondary to pooling of blood within intravascular spaces.<sup>7,25</sup> The lesion may appear more nodular or demonstrate serpiginous regions of increased T2 signal intensity and may demonstrate extraarticular extent. Some regions of the hemangioma may demonstrate signal that is closer to that of adjacent muscle or fatty tissue, and this may represent fibrous septa or vascular channels.<sup>7,10</sup> Long-standing lesions may result in pressure erosions or remodeling of adjacent bone.<sup>10,14</sup> Following intravenous gadolinium administration, there is robust and heterogeneous enhancement of the lesion. It is not common for these lesions to have an associated effusion.<sup>14</sup> MR angiography is a noninvasive test that may also be considered to evaluate the relationship of the hemangioma to adjacent feeding vessels before surgical intervention. Although conventional MRI demonstrates flow voids related to flowing arterial or venous blood, dynamic contrast enhanced MR angiography can also be used to classify the lesion into either low-flow or high-flow entities. This information would prove useful to the surgeon before excision.<sup>28</sup>

Synovial hemangiomas are grossly classified as pedunculated or diffuse. Most commonly, they have been described as synovial; however, they can be juxtaarticular and intermediate as well.<sup>8</sup> Histologically, they are classified as cavernous, capillary, arteriovenous, or venous, based on the predominant type of vascular channel present. In a review of 20 synovial hemangiomas performed by Devaney et al<sup>9</sup> at the Armed Forces Institute of Pathology, the dominant histologic patterns were cavernous (50%), lobular capillary (25%), arteriovenous (20%), and venous (5%).

Treatment and surgery depend on the extent of the lesion. Arthroscopic resection may be performed in lesions that are well defined or pedunculated. Larger lesions, those with diffuse involvement, or those that demonstrate intra- and extraarticular involvement are removed via open surgery often with wide local excision or total synovectomy.<sup>7,20</sup> Recurrence rates are often much higher in cases of diffuse involvement.<sup>20</sup> Outcomes have also been shown

to improve with early treatment, with worsening outcomes in the adult population.<sup>4</sup>

During surgery on our patient, the mass was found to be invading the bone, and upon exploration, we found several craters and erosions into the bone created by the mass that appeared to harbor microscopic neoplasm. A complete excision would have required resection of a substantial amount of bone. Thus, the mass was excised with excision of the entire posterior synovium. Given the presence of bony erosions, the margins were additionally treated with shaving of the bone surface and open application of liquid nitrogen to eradicate any potential microscopic cells and reduce the risk of local recurrence.

The child was treated with a functional elbow brace for 3 months postoperatively. She healed well and regained full motion in the elbow. She is presently 2 years from surgery and without evidence of local recurrence.

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