September 2010 Sports Medicine Case

Left shoulder and neck pain

Date of Initial Evaluation: 6/16/09
CC: left shoulder and neck pain

HPI: This is 9 year old male who presents for evaluation and treatment of left shoulder and neck pain. The pain started about 4 weeks prior to initial evaluation on 5/15/09 as gradual onset neck pain. He awoke one morning with neck stiffness that progressed throughout the day. He was evaluated by his PMD 3 days later. His mom gave him some Tylenol and the pain decreased in about 3 days. Two weeks later, he was in a pool when another child jumped onto his back causing the pain to return. The pain then radiated to the left upper back. He was taken to his PCP on 06/05/09 who thought it might be a muscle strain in the neck. He continues to hold his neck stiffly but is improving according to mom and patient.

In reviewing his history, there are no unexplained constitutional symptoms including rashes, fevers, weight loss, URI symptoms, throat soreness or gum/teeth disease, night time pain or pain at rest.

PMHx: benign, no major illnesses or hospitalizations
Developmental History: normal, full term delivery without complications
PSHx: hypospadius repair at five months of age and a revision in 2008
Immunizations: UTD
Meds: none
NKDA

Food allergies: Watermelon causes hives

Family History: 6 and 7 yo brothers healthy, mother and father healthy, no significant cancer history except the paternal great grandfather was diagnosed with spine cancer in his 70's. Paternal grandfather with hypertension and hypercholesterolemia.
Social Hx: 3rd grade student, lives with both parents. Mom is a homemaker and the father works as a mason.

Physical Exam:
Temperature 98.6 °F (37 °C), temperature source Oral, height 1.406 m (4' 7.35"), weight 50.1 kg (110 lb 7.2 oz).
Constitutional: Well developed, Well nourished, No acute distress and Alert and oriented to person, place, and time
HEENT: normocephalic/ataumatic, anicteric, no LAD
Neck: midline trachea
Tenderness: Yes, left Trapezius muscle, right and left side of neck around C-3 L>R.
Neck skin- no redness, warmth, rashes
Cervical ROM: limited extension, rotation to the left and side bending to the left, full flexion and rotation to the right. As he sits looking forward, his head is slightly turned to the right past midline.

Respiratory: Normal effort, no respiratory distress, no cyanosis
Neurologic: alert, oriented x 3, normal coordination
Psychologic: normal affect, mood and age-appropriate judgment
Skin: Skin over both upper and lower extremities clean, dry, intact

Spine:

No evidence of scoliosis, no cutaneous findings over spine, no obvious rib hump with forward bending.

Neurological:

Upper extremity motor strength and active range of motion at the shoulder, wrist and elbow is normal
Light touch sensation over both upper extremities is normal
There is brisk cap refill distally with normal distal pulses
Gait: reciprocal with no presence of antalgia, no balance deficits

RADIOGRAPHS:

Date: 6/16/09
Views: neck, AP/Lateral/flexion/Extension
My interpretation: C3 vertebral body has ill defined superior border. There appears to be a lucency in the C3 vertebral body. This may represent vertebral collapse. This may represent a lytic process.
DDX:

Bone tumor vertebral body (Chordoma, Giant Cell Carcinoma, Multiple myeloma, hemangioma, metastatic neuroblastoma)
Ewing sarcoma
Bone Lesion- Histiocytosis X, aneurysmal bone cyst
Infectious process (osteomyelitis, TB)
Leukemia with vertebral body collapse

Radiologist’s report: There is minimal prevertebral soft tissue prominence of the upper cervical spine. There is marked compression deformity of C3 with areas of lucency seen in the area of the expected vertebral body of C3 suggestive of a lytic process. There is also some sclerotic abnormal area seen involving the facet region of the C3. There is associated prevertebral soft tissue prominence. Diagnostic considerations include either a neoplastic process such as a histiocytosis versus an infectious process. There is no ligamentous laxity. Visualized upper thorax is normal. Question enlarged sella. An MRI is recommended for further evaluation.

After x-rays done in satellite office of academic center, phone discussion with radiology arranged for urgency MRI via admission through emergency department.

Case update July 2010:

6/16/09 MRI done in Peds ED. Radiologist report: The C3 vertebral body is completely collapsed (vertebra plana), with expansion and surrounding enhancing soft tissue, extending more to the right than the left of the vertebra. There is involvement of the posterior elements, specifically the right lateral mass and transverse process. The intravertebral discs appear preserved in height and signal and do not demonstrate contrast enhancement. The expansion bulges into the posteriorly into the epidural space and anteriorly into the prevertebral space. There is also associated prevertebral soft tissue swelling. There is a mild, focal reversal of the cervical lordosis at the level of the abnormality.

C3 vertebra plana with expansion and soft tissue enhancement. The most likely diagnosis is Langerhans cell histiocytosis/eosinophilic granuloma. Other much less likely differential considerations are primary and metastatic bone neoplasms. Tuberculosis can also affect a vertebral body with sparing of the discs, but is unlikely to cause this appearance.
6/16/09-6/18/09 Admitted to hospital for evaluation and ortho consultation. Bone scan showed mild nonspecific uptake in mid cervical region without focal tracer activity. He was placed in hard collar cervical cast to stabilized cervical spine

7/2009. Underwent CT guided biopsy (pictures enclosed), confirming diagnosis of eosinophilic granuloma. Subsequent evaluation with Oncology. Decision to use oral Methotrexate and Prednisone for 6 months plus Bactrim prophylaxis
8/2009 Ortho follow up transitioned to hard cervical collar. Repeat MRI 8/5/09 showed near resolution of soft tissue swelling around C3. Bone appearance was unchanged.

9/23/2009, 12/16/09 and 3/10/10 Repeat MRIs resolution of soft tissue swelling without significant bone changes

7/8/10 repeat x-rays (pictures below) show persistent vertebral plana. Weaned from brace after 6 months. Clinically he is asymptomatic. Prednisone caused a buffalo hump that is resolving. No complications from methotrexate.

Final Diagnosis: Eosinophilic Granuloma

Discussion:

Pediatric neck pain is a relatively uncommon musculoskeletal complaint. Pediatric spinal cord tumors are relatively rare, accounting for only 1-10% of all central nervous system tumors. Tumor classification is based on anatomic location (extradural, intradural/extradural, intradural/intramedullary). Anatomic location will affect symptom presentation. In one large review, pain (20/35 –57%), weakness (16/35 -46%) and gait disturbance (14/35 – 40%) were the most common symptoms (1). Infection (osteomyelitis, TB), bone lesions (aneurysmal bone cyst, Histiocytosis X,) and insufficiency fractures (primary or secondary osteopenia, metastatic disease) can mimic tumor presentation. A history of persistent pain, unexplained constitutional symptoms including night time pain, upper extremity weakness, dysphagia, motor regression, gait disturbance, limitations in cervical range of motion including new onset torticollis (2) and sensory disturbance should prompt further evaluation. In this case, pain of 4 weeks duration, limitation of cervical range of motion and pain following minor trauma (concern for insufficiency fracture) were all concerning presenting features in this case of a cervical spine eosinophilic granuloma.
Eosinophilic granuloma is the most benign subtype of Langerhans Cell Histiocytosis (LCH). It is a self-limited condition of abnormally proliferating histiocytes from the reticuloendothelial system which causes focal destruction within the bone (3). Most affected patients (90%) are between 5 and 15 years of age. More than 50% of cases involve the skull, spine, pelvis, ribs and mandible. Long bone involvement is typically in the diaphyses of the femur, tibia and humerus. Lesions in the hands and feet are rare. Multiple lesions are demonstrated in approximately 10% of cases at the time of presentation. Vertebral involvement occurs in 6-25% of reported cases. In pediatric patients thoracic spine (54%) and lumbar (35%) involvement are more common than cervical involvement (11%) (4).

The classic radiographic finding of vertebra plana is seen as a complete collapse of the vertebral body on the lateral view. Bone scans are usually negative. MRI often reveals a flare reaction on T2-weighted images, which may be mistaken for a malignant lesion. Differential diagnosis includes Ewing sarcoma, aneurysmal bone cyst, infection, tuberculosis, leukemia, and neuroblastoma (3). MRI findings of well-marginated borders, maintenance of adjacent intervertebral disc spaces, and absence of soft tissue extension are the most important elements in distinguishing eosinophilic granuloma from other causes in the differential diagnosis (5). Because radiographic findings are not pathognomonic, biopsy is usually necessary.

Treatment for isolated spinal lesions includes immobilization and observation (6). Neurological symptoms and spinal (especially cervical) instability may necessitate surgical resection and stabilization. Since most individuals with 4-5 years of growth remaining will remodel the vertebral elements, surgical intervention is not necessary for isolated vertebral collapse (7). Radiation has future risks of malignant transformation and is currently not standard of care (3,4). Chemotherapy is used as primary treatment for widespread disease and may be used to shrink the mass if neurological symptoms are present (6).

In this case, his long term outcome, as with almost all patients of isolated eosinophilic granuloma, is expected to be excellent. He is now asymptomatic. Since he was 9 at age of presentation and has many years of growth remaining, it is expected that he will regenerate at least 2/3 of his vertebral body height. He is currently restricted from contact/collision activities.

References