

April 2011: Radiology Case Study—C2 Chordoma

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DIAGNOSIS: Cervical Chordoma

HISTORY OF PRESENT ILLNESS: The patient is a 13-year-old female who presents for evaluation of posterior neck pain that started 4-5 months ago. The patient denies any precipitating event or injury; however, she is very active in gymnastics and also has a trampoline at home. She describes the pain as an intermittent stiffness and achiness in her posterior neck. The pain was often worse at night. This pain is usually relieved by naproxen which she has been taking regularly over the last few months. The patient's mother reports that she had an initial evaluation with a physical therapist about a month ago, who did not believe that her neck pain was musculoskeletal in nature. The patient's pain has worsened over the past two to three weeks, which caused the patient's parents a lot of concern. X-rays of the cervical spine were obtained and were reportedly normal. The patient continued to experience neck pain daily and subsequently had an MRI of the cervical spine which demonstrated lesions involving C2, C3, and C4. The patient was then referred to the pediatric orthopedic clinic for further evaluation. The patient denies any numbness, tingling, or weakness in the upper or lower extremities. She denies any bowel or bladder incontinence. She reports that the pain has never awoken her from sleep and she has remained very active in gymnastics over the past year despite the neck pain.

PAST MEDICAL HISTORY: Noncontributory, Healthy 13 year old female

PAST SURGICAL HISTORY: Tonsillectomy and PE tube placement for frequent otitis media

MEDICATIONS: Naproxen p.r.n. pain.

ALLERGIES: Penicillin

REVIEW OF SYSTEMS: Negative for frequent cold or sore throat, asthma, pneumonia, recurrent cough, heart murmur or heart problems, seizures or neurologic problems, problems with urination, bedwetting, kidney disease, anemia, diarrhea, constipation, diabetes, metabolic disease, and bowel or bladder incontinence. The patient wears glasses and contacts and has a history of frequent ear infections.

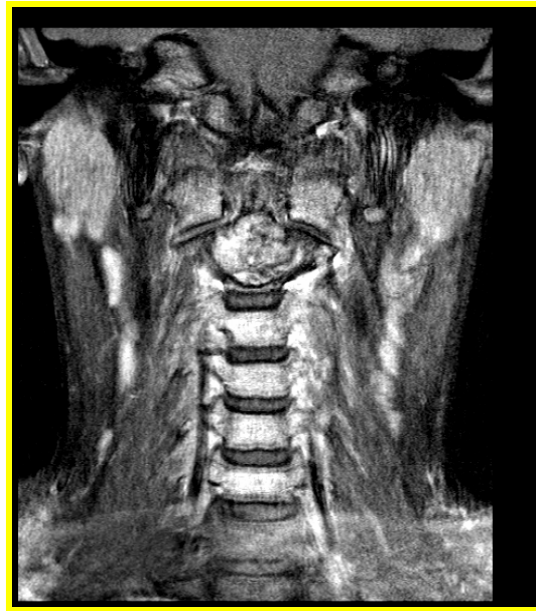
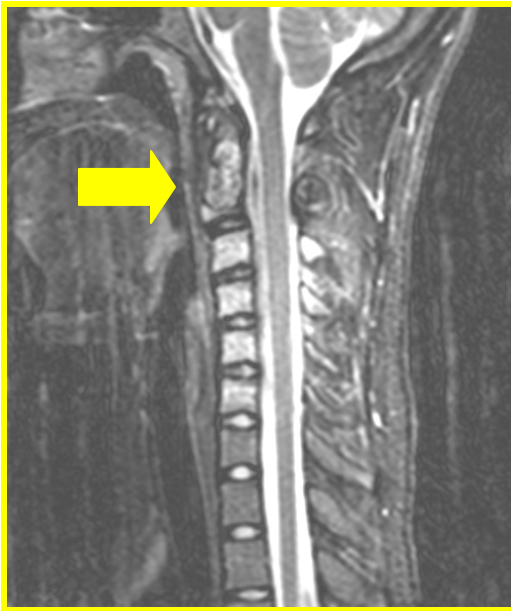
PHYSICAL EXAMINATION: The patient is an alert, conversant, and quiet 13-year-old female in no acute distress. Height 156.5 cm and weight 106.6 pounds. HEENT: Normocephalic and atraumatic. Head with pupils round, equal, and reactive to light and accommodation.

Extraocular motions are intact. Cranial nerves II through XII are intact. Neck: The patient has full range of motion on flexion and extension as well as lateral bending. She has decreased lateral rotation to the left when compared to the right. Rotation to the left also elicits some mild posterior neck pain. There is no step-off to palpation of the posterior elements. Chest: Lungs are clear to auscultation throughout both lung fields. Heart: Regular rate and rhythm without detectable murmurs. Abdomen: Soft, nontender, and nondistended without hepatosplenomegaly or masses. No rebound tenderness. Normal superficial abdominal reflexes. She can toe and heel walk without difficulty. Spine: Straight. No evidence of scoliosis or kyphosis. The patient is nontender to palpation over any aspect of the cervical, thoracic, or lumbar spine. She has no pain with flexion or extension of the spine. Pelvis appears level.

There is no apparent leg length discrepancy. Neurological: Lower extremity neurological examination demonstrates hip flexors, extensors, abductors, adductors, quadriceps, hamstring, EHL, tibialis anterior, FHL, and peronea are 5/5 bilaterally. Sensation is intact from L2 through S2 bilaterally. Deep tendon reflexes of the knees are 2+ and the ankles are 2+. The patient has downgoing toes to plantar reflexes response. Coordination is good and balance is good. Upper extremity neurological examination demonstrates internal and external rotators of the shoulder,

biceps, triceps, brachioradialis, interossei, and lumbricals to be 5/5 bilaterally. Sensation is intact to light touch from C4 through T1 bilaterally. Reflexes are 2+ at the biceps, triceps, and brachioradialis bilaterally. Hoffman is negative bilaterally. Coordination is good by rapid alternating movements.

RADIOGRAPHS: AP/LAT x-rays of the cervical spine show no abnormality, normal alignment, no evidence of fracture. However review of MRI of the cervical spine taken 3 weeks ago demonstrates an expansile lesion involving the C2 vertebral body and posterior elements as well as a lobulated mass-like structure in the vertebral body of C3. These lesions appear lytic and non-aggressive. It was felt that further radiographic studies were needed for complete evaluation and to rule out a more aggressive or malignant process. Therefore a CT was obtained for further evaluation. The CT scan again showed several expansile lytic lesions in the cervical spine



IMPRESSION AND PLAN: The patient is a 13-year-old female who presented to the pediatric orthopaedic clinic with four months of neck pain and stiffness. Review of MRI and CT scan showed multiple lesions in C2, C3, and C4. The differential diagnosis included aneurysmal bone cyst (ABC), hemangioma, Langerhans cell histiocytosis, osteoblastoma, or brown tumor. After the CT scan, a biopsy was recommended. The following week, a CT guided biopsy was done on the C2 lesion. The final diagnosis (final pathology after biopsy): Cervical Chordoma. Cervical Chordomas are very rare tumors that are believed to arise from embryonic notochordal remnants. Chordomas are characterized by slow growth, with local destruction of the bone and extension into the adjacent soft tissue. Distant metastases are occasionally encountered. These tumors usually have a relatively indolent but prolonged course with multiple local recurrences. The treatment generally involves radical surgical excision (which can be very difficult) combined with radiation therapy.