(iv) Primary bone tumours of the growing spine☆

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Abstract

Primary bone tumours of the growing spine are rare and most are benign. However there is a potential for local aggressiveness and malignancies do occur. Surgical resection is the mainstay of treatment and is usually very challenging due to the complexity of local anatomy and proximity to neurovascular structures. In this review we discuss the overall approach to primary bone tumours of the growing spine, focussing on the most common subtypes of benign and malignant tumours seen in this subset.

Keywords aneurysmal bone cyst; eosinophilic granuloma; Ewing's sarcoma; growing spine; Langerhans cell histiocytosis; osteoblastoma; osteogenic sarcoma; osteoid osteoma; osteosarcoma; spine tumours

Introduction

Primary bone tumours of the growing spine are rare, accounting for less than 5% of all primary bone tumours.[1] Approximately 70% of bone tumours in children and adolescents are benign,[2] however malignant tumours may occur and are extremely difficult to manage. Progress in imaging, such as high resolution magnetic resonance imaging (MRI) and computed tomography (CT); improved surgical and spinal cord monitoring techniques; and the advances in medical oncology have improved the ability to make the correct diagnosis, and outcome of children with bone tumours of the growing spine.

Statistics from the surveillance, epidemiology, and end results program (SEER) of the National Cancer Institute reveal that about 1000 children and adolescents under the age of 20 are diagnosed with primary bone malignancies in the United States each year. Of those approximately 50% of children and adolescents have osteosarcoma and 30% have Ewing's sarcoma.[3] These malignancies represent 6% of all childhood cancers and spine tumours comprise only a small part of this total.

Pain is the most common presenting symptom of children with bone tumours of the spine in approximately 90%.[4] Compared to the usual evaluation and treatment for back pain in adults, children with significant back pain deserve earlier and more extensive work-up, since far less is known about the true incidence and causes of back pain in this age group. Some estimate the prevalence of back pain in the 2nd decade as high as 36%,[5] with only 10% or less of these children requiring or seeking medical assistance.[5] The most frequent causes of back pain in children are trauma, infection, spondylolysis and spondylolisthesis.

When a child presents for evaluation of back pain and a bone lesion is identified, there is often anxiety on the part of the family and evaluating physician. Nonetheless, bone tumours are far less common than fractures, normal developmental variations, congenital malformations, infection, metabolic disorders, and tumour-like lesions. The clinical and radiographic features of these groups however may overlap.

During the evaluation and management of bone tumours of the growing spine, several steps and principles should be followed:

- Complete clinical work-up with high-quality and appropriate imaging, for adequate tumour staging.
- When a biopsy is performed, the technique used should be simple and safe, allowing for the best diagnostic yield without interfering with the definitive surgical plan.
- When indicated, adjuvant treatments or modalities should be used based on tumour natural history, stage, and the child's overall condition.
- If surgery is indicated, it should be performed based on a standardized spinal surgical staging system and the goals should be set and delineated in regards to margins and outcome.
- Whenever possible, efforts should be made to preserve neurological function, spine alignment and stability.

In this review we will focus on the general approach to bone tumours of the growing spine and discuss in further detail the current state of the art of the most common benign and malignant tumours of the spine in this age group.

Clinical presentation

Persistent back pain either generalized or localized to a specific area of the spine is the most common presenting symptom in children with spine tumours. Tumour pain is often unrelenting, progressive, and usually present during the night, disturbing the child's sleep. Firm palpation or percussion over the involved area can usually reproduce the pain. A palpable mass is rarely found on exam. The pain is usually caused by tumour growth with subsequent expansion and/or deformity of the vertebral body. Sometimes it is associated with pathologic fractures (at times leading to spinal instability), and invasion of the paravertebral soft tissues. In general, back pain in children should be taken seriously, often warranting further investigation. Contrary to the natural course seen with benign tumours, pain from malignant tumours usually increases in intensity quite rapidly and can become debilitating in the course of just a few weeks. Unfortunately, pain may not be well expressed by young children, and a change in their personality or the presence of constitutional

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symptoms such as lethargy, or signs such as fever or limping may be initial indicators. Malignant tumours, particularly Ewing sarcoma, may also present with constitutional symptoms such as fever, malaise, and weight loss.

Spinal deformities may sometimes be the only presenting symptom, especially with benign tumours. Atypical scoliosis may develop in patients especially with osteoid osteoma or osteoblastoma of the spine.[6]

Although uncommon, neurological compromise at presentation may occur and is usually a result of one of the following mechanisms: extrinsic compression of nerve roots by the tumour; extrinsic compression of nerve roots as a result of a pathologic fracture; extrinsic compression of vascular structures that supply the spinal cord or nerves; and direct tumour invasion of nerves. Clinical presentation will depend on the level of involvement, for example compression at the level of the conus medullaris can lead to sphincter dysfunction.

Imaging

Radiographs should be part of the initial assessment whenever a neoplasm is suspected, often providing valuable information regarding the nature and behaviour of the lesion. Three-foot posterior–anterior and lateral radiographs are recommended to evaluate scoliosis or sagittal plane deformity. Vertebral collapse (due to pathologic fracture) is a common radiographic finding associated with benign–aggressive and malignant spine tumours. Radiographs help to differentiate between discitis/osteomyelitis and tumour, since infection usually starts at the vertebral end plate (due to the rich blood supply of the area) and results in disc destruction and narrowing, while the intervertebral disc is very resistant to tumour invasion. Posterior element lesions may produce the "winking owl" sign due to obliteration of one of the pedicles in the antero-posterior view.

CT scan is a noninvasive and quick exam that allows a 3-D view of the bone architecture in the spine, as well as screening for metastatic lung disease. CT is generally well tolerated by younger patients but there are increasing concerns about potentially high levels of radiation exposure. Bone windows can often help differentiate between benign, slow-growing lesions and malignant lesions. CT is helpful to evaluate bone architecture and stability and is often used to guide needle biopsies in areas of difficult surgical access. 3-D reconstruction images can be used to build spine models used to plan complex resections.

MRI plays an essential role in the evaluation of tumours of the growing spine, and helps determine the exact location and size of the lesion, evaluate the presence of an associated soft-tissue mass, differentiate soft tissue from fat, fluid, or haemorrhage, and delineate the relationship between tumour and spinal cord, cauda equina, nerve roots, and other neurovascular structures. Other advantages include being noninvasive, painless, and without radiation. Disadvantages include the usual need for sedation and occasionally general anaesthesia for infants and young children. MRI is also used to evaluate for local recurrence, and the use of titanium instrumentation systems (when stability is at risk) allows high-quality MR images to be obtained with minimum scatter.[7]

Isotope bone scans are used for identification of occult lesions in children with persistent back pain and/or atypical scoliosis. It is sensitive in localizing accurately small pathologic processes involving the spine, such as osteoid osteoma. Bone scanning is also important in the staging of bone tumours to rule out multiple lesions, such as with Langerhans cell histiocytosis or metastatic disease. Newer imaging techniques, such as single photon emission computed tomography (SPECT) and positron emission tomography (PET) also provide information regarding grading, therapy response or recurrence.

Biopsy

Prompt and appropriate treatment of tumours of the growing spine depends on an accurate diagnosis; so biopsy is an essential step. Complications as a result of an inappropriate biopsy, altering negatively the definitive management, are seen in approximately 20% of patients with extremity tumours.[8] It is likely that this rate is even higher for patients with spine lesions. The biopsy should preferably be done by an experienced orthopaedic tumour surgeon, at a tertiary centre that routinely cares for children with bone tumours.

There are two types of biopsies used for spine tumours. Percutaneous or needle biopsies are often done *via* CT or fluoroscopic guidance. The main advantage is the less-invasive nature of the procedure and speedy recovery. The main disadvantage is the possibility of inadequate tissue sampling (quantity) for the multiple diagnostic tests required (routine haematoxylin and eosin, immunohistochemistry, molecular genetic studies, and other research samples). Percutaneous techniques are generally preferred for lesions in locations where surgical access is difficult, and for lesions with a narrow differential diagnosis.

Open incisional biopsy has the advantage of adequate tissue sampling (quantity), and less chance of needing a 2nd biopsy. Also, for benign tumours, an open biopsy may be followed by excision of the lesion in the same anaesthetic event. The main disadvantages are potential blood loss, increased morbidity, tissue contamination and longer recovery. Thoracoscopy for thoracic spine biopsy (mainly anterior elements) may be appropriate in selected instances, but only if the risk of contamination of the thoracic cavity with the procedure is not a concern.

Treatment overview

Clinical staging systems aim to describe the behaviour, management principles and prognosis of a given tumour. They are usually based on the tumour characteristics such as type, size, and grade, and the clinical extension of the disease (i.e. presence of metastasis). Surgical staging systems, especially about the spine, aim to guide local surgical control and strategies, assisting in the pre-operative planning of complex resections. Among the most used systems are the Weinstein–Boriani–Biagini system (WBB) and the Tomita system. The staging is done based on imaging findings on CT, MRI and sometimes angiography.

The WBB system divides the vertebrae in the transverse plane into 12 radial segments (such as in a clock's face) and into five layers (from paravertebral extraosseous region to the duramater).[9] The Tomita system assigns numbers from 1 to 5 based on tumour location within the vertebra (1 = body; 2 = pedicles; 3 = posterior elements; 4 = spinal canal; 5 = intervertebral space), and from 1 to 7 based on the number of affected areas plus compartmental involvement (intra *vs.* extra-compartmental).[10]