Common neoplastic causes of paediatric and adolescent back pain

Abstract

Neoplasm of the spinal column in children is rare, but can involve either benign or malignant tumours. Early detection of malignant tumours is key to successful clinical outcome and long-term prognosis. In such cases, back pain is a common presenting symptom, but often has a non-neoplastic cause. Therefore, it is important for GPs and trainees who encounter paediatric patients to be aware of the clinical entity to be able to thoroughly assess them in clinical practice. This article discusses the types of paediatric spinal neoplasms, anatomical-based classification, clinical red flags, imaging modalities and outlines brief management options.

Key words: Back; Epidemiology; Investigation; Management; Neoplasm; Paediatric; Pain; Prognosis; Spinal

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Background

Epidemiology of paediatric back pain

Paediatric back pain is increasing in prevalence, with a systematic review of 27 studies quoting lifetime prevalence at 40% and annual incidence at 15% (Kamper et al, 2016). This review found that prevalence is low in children under the age of 12 years but increases quickly after this age, with adolescent prevalence almost matching that in adults.

The majority of cases of back pain in children and adolescents are diagnosed as mechanical back pain, or do not receive a definitive diagnosis in up to 50% of cases (Ryder et al, 2015). Despite this, it can still be the initial presentation of serious pathology, including neoplasia (both benign and malignant), with one study reporting that 34% of children who present with to a paediatric orthopaedic clinic with back pain will have an identifiable pathology (Ramirez et al, 2015). Children often present to primary care providers, so a systematic approach to history and examination with a knowledge of red flag features of back pain is essential to select the best investigations (Tomlinson and Gummerson, 2017).

Epidemiology of back pain caused by neoplastic processes

It is estimated that 40 children are diagnosed with a tumour involving the spine or spinal cord every year in the UK (Wilne et al, 2010). Although spine and spinal cord tumours only account for 2% of childhood neoplasia, these tumours are associated with a disproportionate degree of morbidity. Diagnosis is often delayed in children, meaning that they are frequently symptomatic for months beforehand, and many have developed disabling neurological deficits at the time of diagnosis, which are often only partially reversible.

Understanding common neoplastic causes of paediatric and adolescent back pain

It is important to understand the neoplastic causes of back pain to ensure appropriate investigation of children presenting with back pain. This will allow timely treatment, and avoid over-investigation of children with non-specific musculoskeletal pain. Knowledge of the common neoplastic causes of back pain can help the clinician to make the correct diagnosis and provide appropriate treatment.

This review will facilitate earlier diagnosis of spinal neoplasms in children, thus reducing the degree of permanent disabling neurological deficit. This article will:

- Review the current literature on paediatric back pain and spinal neoplasia
- Outline serious spinal pathology

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- Describe common presentations of the various causes
- Highlight key features in the assessment of paediatric back pain
- Briefly describe imaging and management
- Suggest ways this assessment can be improved in the future.

Causes

Pathology and classification of tumours

Tumours are classified based on where they appear in the spine, so accurate classification requires an understanding of spinal anatomy. The anatomy of the spine and spinal cord is outlined in Figure 1.

Spinal tumours can be either primary or secondary. Primary neoplasms only account for around 10% of all spinal neoplasms among people in all age groups (Harshavardhana and Dormans, 2018). In people under the age of 20 years, the majority of spinal neoplasms are benign and haematological malignancies account for the majority of metastatic lesions.

Spinal tumours can be divided into those that arise external to the meninges (extradural) and those within (intradural). Intradural tumours can be further divided into intramedullary and extramedullary (Wilne et al, 2010). Classification of spinal tumours is shown in Figure 2 (Baruah et al, 2015; Garcia et al, 2015).

Extradural tumours can be divided into malignant or benign, as shown in Figure 2. Malignant bone tumours in children account for around 5% of all malignant neoplasms, with the majority being Ewing's sarcomas or osteosarcomas (Egea-Gámez et al, 2019).

Clinical picture

Clinical presentation and assessment

Back pain is a leading complaint of children with spinal neoplasms, with 25–30% of children complaining of recurrent episodes of back pain (Huisman, 2009). Back pain can be classified as spinal, root or tract pain. The majority of cases present with spinal pain,

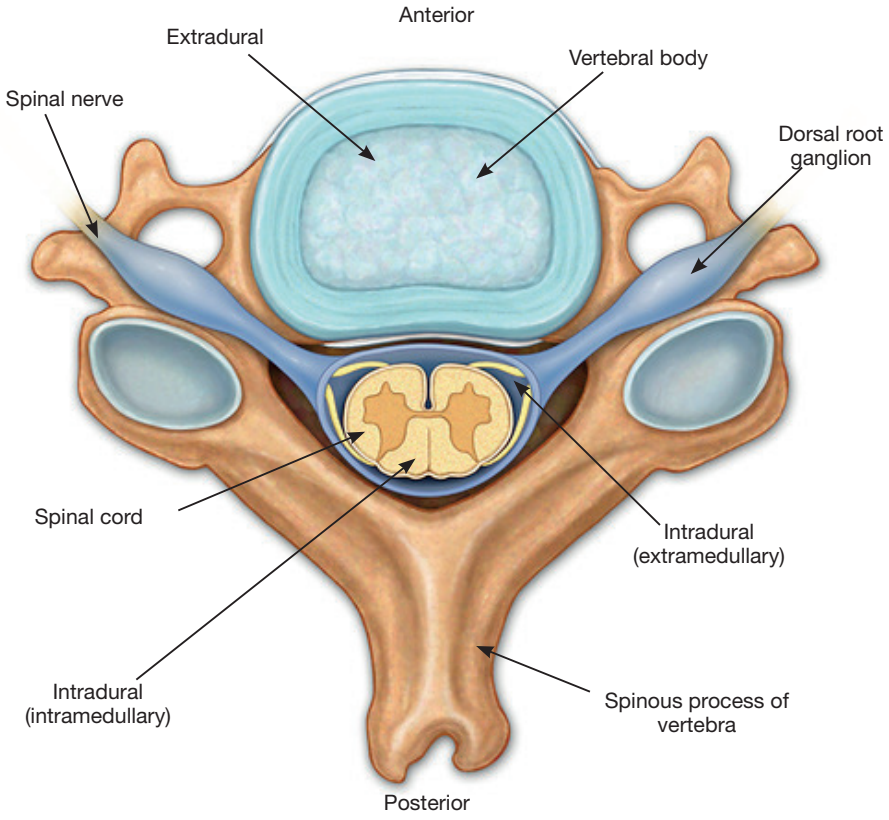

Figure 1. Anatomy of spinal cord and spine.

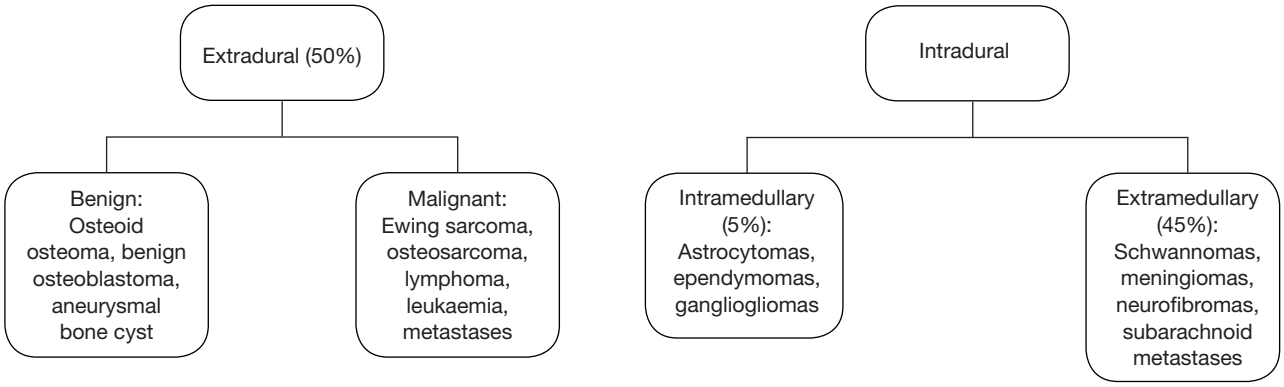

Figure 2. Classification of spinal tumours.

classically a dull ache in nature, and present in the vertebral segments adjacent to the tumour. Root pain mimics that of disk herniation, and tract pain is a burning sensation, usually as the result of the tumour infiltrating the spino-thalamic tracts within the spinal cord itself.

Certain features in the history and examination of a child or adolescent with back pain should immediately warrant further investigation, such as pain lasting more than a month, weight loss, neurological deficits, immunosuppression or rheumatological disease, as these point to potentially sinister underlying pathology (Freeman and Govindarajan, 2012). **Table 1** outlines red flag features in the assessment of a child with back pain (Ryder et al, 2015).

Specific clinical features can also be characteristic of certain pathologies. For example, malignant lesions are more likely to have acute, severe pain, with rapid onset of symptoms and pathological fractures, whereas benign pathologies are more likely to be chronic and indolent, with mild to moderate pain. In addition, night pain can be characteristic of osteoid osteoma or osteoblastoma, with the night pain of osteoblastoma characteristically improving upon use of non-steroidal anti-inflammatories in up to 70% of cases (Harshavardhana and Dormans, 2018).

Some spinal neoplasms can be associated with other conditions. One example is neurofibromatosis type 2 and the development of ependymomas and schwannomas (Furtado et al, 2016). Another is von Hippel–Lindau syndrome and haemangioblastomas, with approximately 30% of all haemangioblastoma patients having this syndrome (Hazenfield and Gaskill-Shipley, 2016). Features of the above syndromes in association with back pain or other red flag features should increase the suspicion of a spinal tumour.

A thorough examination of the spine is an essential component of the assessment of paediatric back pain. Younger children may have to be examined supine, to aid compliance and therefore accurately assess alignment and wasting, whereas older children can be examined standing (Aresti and Barry, 2016). Examining the spine should begin using

Table 1. Red flag features suggestive of significant pathology

History of previous malignancy
Exposure to tuberculosis
Immunosuppression
Night pain (especially if causing to awaken)
Early morning stiffness >45 minutes
History of significant trauma
Age <5 years
Peripheral neurological abnormalities
Bladder or bowel dysfunction

the classic 'look, feel, move, special test' approach. As with the history, specific signs can point to specific diagnoses, such as painful secondary scoliosis and osteoid osteoma (Ciftdemir et al, 2016). As well as this, a thorough neurological examination, including a rectal examination, must be undertaken to elicit any signs that point towards more sinister pathology, as neurological deficit itself is a red flag (Rossiter et al, 2017).

Imaging

X-rays are often used as first-line imaging investigations because of the ready availability and low cost (Lateef and Patel, 2009). If X-rays are negative but the clinician has a high index of suspicion because of red flag features such as constant, night or radicular pain and/or abnormal neurological examination, magnetic resonance imaging scans are normally the next investigation required (Feldman et al, 2006). The diagnostic yield can also increase; magnetic resonance imaging established an identifiable diagnosis in an additional 25% of patients (Ramirez et al, 2015). Magnetic resonance imaging scans also accurately assess the distribution, features, localisation and extent of spinal cord tumours, helping guide management (Chowdhary et al, 2017).

Using these two imaging modalities brings a high yield of diagnoses of back pain, with one study finding that of 31 patients found to have a diagnosis, 68% were diagnosed from X-rays and the remaining 32% from magnetic resonance imaging scans, all of which had negative X-rays but red flag features (Feldman et al, 2006).

Table 2 describes the indications, advantages and disadvantages of different imaging modalities used in the assessment of paediatric back pain (Lateef and Patel, 2009; Aresti and Barry, 2016; Tomlinson and Gummerson, 2017; Harshavardhana and Dormans, 2018).

Management

The mainstay of management of spinal neoplasms is surgical resection, as this is the most effective way to relieve symptoms (Atkinson et al, 2016). Some tumours, such as osteoid osteomas, can be treated with surgical resection alone. Neo-adjuvant chemotherapy followed by wide resection is the treatment of choice for Ewing's sarcomas because of the high cellularity of the tumour tissue and in osteosarcomas, which also receive postoperative chemotherapy. Other tumours such as osteoblastomas benefit from postoperative radiotherapy for local control (Ciftdemir et al, 2016). All of the above will require a multidisciplinary approach, taking into account clinical and magnetic resonance imaging features to guide approach and management.

Prognosis

Prognosis of paediatric spinal tumours depends on a number of factors, including the type of tumour. Ewing's sarcoma and osteosarcoma traditionally carry a worse prognosis because tumours arising from the vertebrae often cannot be fully resected (Wilne et al, 2010). However,

Table 2. Imaging modalities in assessment of paediatric back pain

Type of imaging	Indication	Advantages	Disadvantages
Plain X-ray	■ Clinical suspicion ■ Red flag features	■ No need for sedation ■ Quick ■ High availability and low cost	■ Can be negative in early stages ■ Radiation exposure ■ Cannot visualise discs
Magnetic resonance imaging	■ Abnormal neurology ■ Negative X-ray findings with red flag features	■ Sensitive for infection and tumours ■ Provides accurate staging ■ No radiation exposure ■ Better visualisation of soft tissue	■ Potential need for sedation in younger children
Computed tomography	■ Demarcation of bony anatomy	■ Bony architecture visualised more readily, especially cortical bone	■ Large dose of radiation

Key points

- Diagnosis of spinal neoplasia in children is often delayed, leading to a higher morbidity rate.
- Around 25–30% of children with a spinal neoplasm present with recurrent back pain.
- A thorough search of red flag symptoms and abnormal signs on examination enables clinicians to refer or reassure children presenting with back pain.
- Magnetic resonance imaging scans should be considered in a child with a negative X-ray but high clinical suspicion.
- The main management of spinal neoplasms is surgical resection, with some patients requiring neoadjuvant chemotherapy or radiotherapy.
- The 5-year survival rate of spinal neoplasms has increased from 10% to 70% in the last 40 years.

the 5-year survival rates of these has increased from 10% before the 1970s to 70–80% in recent years as a result of the development of adjuvant therapies (Ciftdemir et al, 2016).

Those who present with significant preoperative motor deficit are unlikely to show much improvement, highlighting the importance of picking up the diagnosis early before it gets to this stage (Wilne et al, 2010). Intramedullary tumours also carry a worse prognosis for functional improvement. In one study looking at long-term outcomes in children with intramedullary tumours, functional improvement was noted in 55% of patients by last follow up (Ahmed et al, 2014).

Conclusions

Thorough clinical evaluation of all paediatric back pain is essential, as is a clear understanding of red flags to avoid misdiagnosis or delayed diagnosis of potential malignant pathology. Early detection, with modern surgical techniques and neoadjuvant chemotherapy or radiotherapy, can allow successful management of malignant neoplasms around the spinal column.

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Conflicts of interest

The authors declare no conflicts of interest.

References

Ahmed R, Menezes AH, Awe OO, Torner JC. Long-term disease and neurological outcomes in patients with pediatric intramedullary spinal cord tumors. J Neurosurg Pediatr. 2014;13(6):600–612. https://doi.org/10.3171/2014.1.PEDS13316

Aresti NA, Barry M. Paediatric spine. In: Aresti N, Ramachandran M, Paterson M, Barry M, eds. Paediatric Orthopaedics in Clinical Practice. London: Springer; 2016:53–68. https://link.springer.com/content/pdf/10.1007%2F978-1-4471-6769-3_5.pdf (accessed 22 February 2020)

Atkinson R, Stephenson J, Mima C et al. An assessment of key risk factors for surgical site infection in patients undergoing surgery for spinal metastases. J Wound Care. 2016;25(9):30–34. https://doi.org/10.12968/jowc.2016.25.Sup9.S30

Baruah D, Chandra T, Bajaj M et al. A simplified algorithm for diagnosis of spinal cord lesions. Curr Probl Diagn Radiol. 2015;44(3):256–266. https://doi.org/10.1067/j.cpradiol.2014.12.004

Chowdhary AS, Durai B, Mohanty J. MRI evaluation of spinal cord tumours with histopathological correlation. J Evid Based Med Healthc. 2017;4(95):5978–5986. https://doi.org/10.18410/jebmh/2017/1206

Ciftdemir M, Kaya M, Selcuk E et al. Tumors of the spine. World J Orthop. 2016;7(2):109. https://doi.org/10.5312/wjo.v7.i2.109

Egea-Gámez RM, Ponz-Lueza V, Cendrero-Torrado A et al. Spinal osteosarcoma in the paediatric age group: case series and literature review. Rev Esp Cir Ortop Traumatol. 2019;63(2):122–131. https://doi.org/10.1016/j.recot.2018.09.001

Feldman DS, Straight JJ, Badra MI, Mohaideen A, Madan SS. Evaluation of an algorithmic approach. J Pediatr Orthop. 2006;26(3):353–357. https://doi.org/10.1097/01.bpo.0000214928.25809.f9

Freeman JD, Govindarajan P. Paediatric back pain and a limp. Emerg Med J. 2012;29(6):523–523. https://doi.org/10.1136/emermed-2011-201063

Furtado AD, Panigrahy A, Fitz CR. CNS and spinal tumors. 1st edn. Pittsburgh: Elsevier B.V; 2016.

Garcia CM, Sandoval Garcia C, Bragg T et al. An 11-year-old boy with a sacral spinal mass. Brain Pathol. 2015;(25):653–654. https://doi.org/10.1111/bpa.12285

Harshavardhana NS, Dormans JP. Tumours of the cervical spine. In: Philips JH, Hedequist DJ, Shah SA, Yaszay B, eds. The management of disorders of the child's cervical spine. Boston (MA): Springer; 2018: 133–154.

Hazenfield JM, Gaskill-Shipley MF. Neoplastic and paraneoplastic involvement of the spinal cord. Semin Ultrasound CT MRI. 2016;37(5):482–497. https://doi.org/10.1053/j.sult.2016.05.009

Huisman T. Pediatric tumors of the spine. Canc Imag. 2009;9(Special Issue A):S45–48. https://doi.org/10.1102/1470-7330.2009.9012

Kamper SJ, Yamato TP, Williams CM. The prevalence, risk factors, prognosis and treatment for back pain in children and adolescents: An overview of systematic reviews. Best Pract Res Clin Rheumatol. 2016;30(6):1021–1036. https://doi.org/10.1016/j.berh.2017.04.003

Lateef H, Patel D. What is the role of imaging in acute low back pain? Curr Rev Musculoskelet Med. 2009;2(2):69–73. https://doi.org/10.1007/s12178-008-9037-0

Ramirez N, Flynn JM, Brian WH et al. Evaluation of a systematic approach to pediatric back pain: the utility of magnetic resonance imaging. J Pediatr Orthop. 2015;35(1):28–32. https://doi.org/10.1097/BPO.0000000000000190

Rossiter DJ, Haider Z, David B et al. How not to miss major spinal pathology in patients with back pain. Br J Hosp Med. 2017;78(5):C66–C69. https://doi.org/10.12968/hmed.2017.78.5.C66

Ryder C, Harrison K, Johnson K. Back pain in children and adolescents. Paediatr Child Health. 2015;25(12):550–554. https://doi.org/10.1016/j.paed.2015.10.002

Tomlinson JE, Gummerson NW. Paediatric spinal conditions. Surgery. 2017;35(1):39–47. https://doi.org/10.1016/j.mpsur.2016.10.013

Wilne S, Wilne S, Walker D. Spine and spinal cord tumours in children: a diagnostic and therapeutic challenge to healthcare systems. Best Pract Arch Dis Child Educ Pr Ed. 2010;95(2):47–54. https://doi.org/10.1136/adc.2008.143214