Occult Spinal Dysraphism: A Challenge in Pain Management

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Spina bifida is a common birth defect affecting the central nervous system and represents a group of neural tube defects caused by congenital dysraphic malformations of the vertebral column and/or spinal cord. The anatomy in these patients is challenging and includes structural and vascular abnormalities including arteriovenous malformation or fistulae, and fatty substitution of paravertebral tissues. A magnetic resonance image (MRI) is needed for management of patients with lumbar radiculopathy and clinical features suspicious of occult spinal dysraphism. Risks and benefits of lumbar epidural steroids should be discussed comprehensively with those patients and in the best case scenario be avoided. Occult spinal dysraphism poses a clinical dilemma for interventional pain specialists managing those patients with lumbar radiculopathy. We report a case of occult spinal dysraphism discovered following the development of post-traumatic radicular symptoms.

Key words: Occult spinal dysraphism, spina bifida, lumbar radiculopathy, pain, pain management, physical findings

Occult spinal dysraphism (OSD) refers to the failure of the vertebral bodies to fuse due to an abnormal union of the posterior vertebral arches with unexposed neural tissue. This abnormal neural tube closure occurs between the third and fourth weeks of gestation and the term spinal dysraphism includes the overall group of defects derived from the maldevelopment of ectodermal, mesodermal, and neuroectodermal tissues, and its sequelae may affect brain, bones, extremities, and bowel and bladder functions. There is strong evidence that there has been a decline in the incidence worldwide since 1970s. The decline is probably due to systematic use of dietary folic acid and, more recently, to the advent of prenatal diagnosis. This congenital condition presents a unique challenge to interventional pain management for such patients who suffer from radicular symptoms. We present a patient with spinal dysraphism who had radicular symptoms following a motor vehicle accident.

Case Report

A 43-year-old man presented to the pain clinic with low back pain radiating down to his right leg. The pain started 5 months ago following a car accident. It was moderate to severe in intensity, constant, sharp, shooting in nature, and radiating down posteriorly to his right leg up to the level of his ankle. It was associ-
ated with tingling and numbness in the right leg. Pain was aggravated with exercise, walking, and excessive movements, and partially relieved with rest and nonsteroidal anti-inflammatory drugs (NSAIDs).

Examination revealed dimpling of the skin with hyperpigmentation in his lower lumbar region (Fig. 1) as well as positive right straight leg raise test. Based on this abnormal physical exam, a lumbar spine magnetic resonance imaging (MRI) was done which showed occult spinal dysraphism with close opposition at L4 level. At this location, there was a tenting of the thecal sac and a tract within the subcutaneous soft tissues extending up to the overlying skin (Fig. 2, Fig. 3). An epidural steroid injection was contraindicated in view of the patient’s anatomic abnormality; therefore, pain was controlled effectively with NSAIDS and opioids. At the patient’s 6 week follow-up examination, he had significant relief of his symptoms.

**Discussion**

Spina bifida is the second most common childhood abnormality/disability disease, following cerebral palsy. It represents a group of neural tube defects caused by congenital dysraphic malformations of the vertebral column and/or spinal cord. It is the most frequent spinal cord disorder in children (1). The highest incidence occurs in the British Islands, Ireland, Wales, and Scotland, and the lowest in Japan. In the U.S., the rate of neural tube defects was about 0.6 per 1,000 births in 1989, with a higher incidence in families of Irish, German, or Hispanic ancestry and lower among Asians and Pacific Islanders. The factors implicated are both inherent and environmental. The incidence is increased slightly in women compared to men (1.2 – 1). Recurrence rate is 2.5 to 5% after the birth of one child with spina bifida and doubles after 2 affected children (2).
Several environmental risk factors have been implicated which include low socioeconomic class, midspring conception, maternal obesity/Diabetes Mellitus, in utero exposure to anticonvulsant drugs (valproic acid, carbamazepine), and maternal febrile illness (hyperthermia). Various studies have demonstrated that folic acid (0.4 mg daily) periconceptually and during early pregnancy significantly reduce the occurrence and recurrence of neural tube deficits (3). Spina Bifida Occulta (Occult Spinal dysraphism [OSD]) affects primarily the vertebrae. The neural and meningeal elements are not herniated. A frequent sign in 50% of children is the presence of a pigmented nevus, angioma, hirsute patch, and dimple or dermal sinus on the overlying skin (4). Spina bifida cystica collectively designates meningocele, myelomeningocele, and other cystic lesions. In spina bifida cystica, the contents of the spinal canal herniate through the posterior vertebral opening.

Patients are often asymptomatic with only minor skin abnormalities. Clinically, they can be difficult to diagnose and are often missed until later in life. Most commonly, they are suspected after detection of associated findings on the skin, which was the case in our patient. Generally, symptoms are due to tethering or stretching and, occasionally, compression of the lower spinal cord. Severe pain in the lower back, radiating into the legs, groin, and perineum could occur as well as bilateral leg weakness, numbness, and loss of feeling in lower extremities. Urinary and/or bowel symptoms may also occur (5). OSD may be associated with other cord abnormalities such as fusion of bony laminae, vertebral anomalies, meningeal hernias, and fatty substitution of paravertebral tissues, which could make the identification of the epidural space more difficult and increase the risk of accidental dural tap if an epidural is attempted. The risk of spinal damage, intra-arachnoid hematoma, and/or spinal infection could be much higher than usual.

Management of patients with lumbar radiculopathy with OSD signs or suspicious findings on physical exam needs an MRI to further delineate the anatomy. Benefit and risks of lumbar epidural steroids should be discussed comprehensively with the patient. There is a paucity of literature and currently there is no current guideline for the management of these patients; however, conservative management with NSAIDs and opioids – for severe pain – should be strongly considered.

**Conclusion**

The goal of this case report is to raise awareness about OSD’s subtle presentation in patients with lumbar radiculopathy. A meticulous examination should be performed on any patient with lower back pain and especially where OSD is suspected. An MRI should be ordered prior to performing an epidural steroid injection or any other interventional procedure since anatomic abnormalities in these patients could have serious neurological consequences. Finally conservative management with NSAIDs and maybe short-term opioids is advisable.

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