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UNUSUAL MALFORMATIONS IN OCCULT SPINAL DYSRAPHISM*

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Two cases of occult spinal dysraphism with different clinical symptoms, signs and congenital pathologies are presented. One had malformations including scoliosis, dermoid tumor, hydromyelia, diastematomyelia, dermal sinus, low conus, vertebrae anomalies and dextrocardia. The occurrence of dextrocardia in association with occult spinal dysraphism was found to be extremely unusual. The second case is presented in relation to the rarity of teratoma with dermal sinus and tethered cord in the lumbar area. Myelography, computed tomography (CT), Myelo CT and magnetic resonance were used in making a diagnosis. *Key words: spinal dysraphism, dermal sinus, diastematomyelia, teratoma, dextrocardia.*

Occult spinal dysraphism is an entity composed of a variety of malformations of embriological development. It typically causes progressive neurologic deterioration, whether of bladder, bowel or lower extremity sensorimotor function. Tethered cord due to lipoma, lipomyelomeningocele, tight filum terminale and adhesion, dermal sinus with or without tumors, diastematomyelia, skin and spine abnormalities have appeared under the term occult spinal dysraphism[1-4]. More than one type of lesion tends to occur in a single case.

The symptoms of these lesions may arise at any age, mostly occurring in the pediatric group[1, 5, 6] and rarely in adulthood and late life[7, 8]. Myelography, myelo computerized tomography (CT) and particularly magnetic resonance imaging (MRI) have made possible the identification of the exact location and have simplified the surgical procedure of these malformations[2, 9-11].

Case Reports

Case 1

A 13-month-old infant was admitted with the complaint of inability to stand or sit straight. Neurologic examination revealed paraparesis, analgesia under L3

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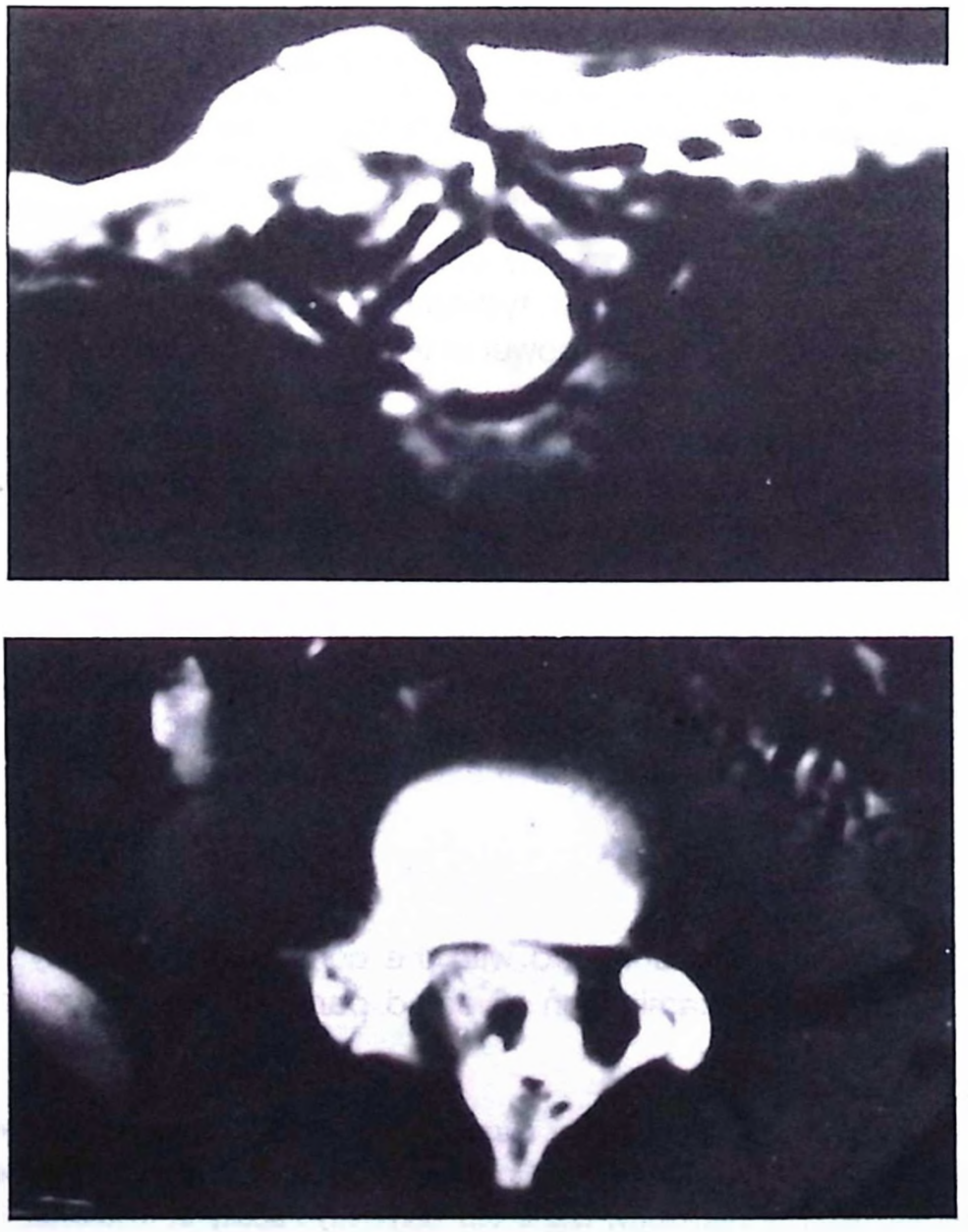
dermatomes, and absent anal and achilles reflexes. In the mid-dorsal area, an outwardly protruding, mobile mass with a diameter of 1 cm was noted. In the mid-sacral region the skin was dark blue and included a dermal sinus.

Plain radiographs demonstrated dextrocardia and vertebrae anomalies between T10 and T12 vertebrae. MR imaging disclosed two masses located in the subarachnoid space and the subcutanous layer and a tract between them at the level of T9-10. The spinal cord lost its normal configuration, had irregular borders and included hydromyelia proximal to the mass. A bony septum at the T11 level and bifid conus at the L5 level were also visualized (Figs. 1a,. b).



Figs. 1a, b: Magnetic Resonance Image of the thoracic and upper lumbar area in the coronal plane. Hydromyelia, tumoral mass and diastematomyelia are clearly visualized.

Complete laminectomy was performed at the T8-12 levels. The masses located in the subcutaneous layer and in the subarachnoid space and the tract between them were removed completely with microtechnique. The bony septum was drilled. At the level of T11, the cord divided in two unequal parts with their own separate pial, arachnoid and dural coverings. The dural tubes reunited below the septum into a single tube, though the split cords continued caudally. While closing the dura, bradycardia and then cardiac arrest occurred. After three hours the patient died. The patient's family refused autopsy.

The tumor had two cystic components and a tract between them. The cysts were composed of matted hair as well as soft, white, cheesy material. Histological sections revealed stratified squamous epithelial cells in the cyst wall and keratinocious lamelles inside. The tract was composed of stratified squamous epithelium in papillary structure with underlying sebaceous glands, hair follicles and capillaries (Fig. 2).

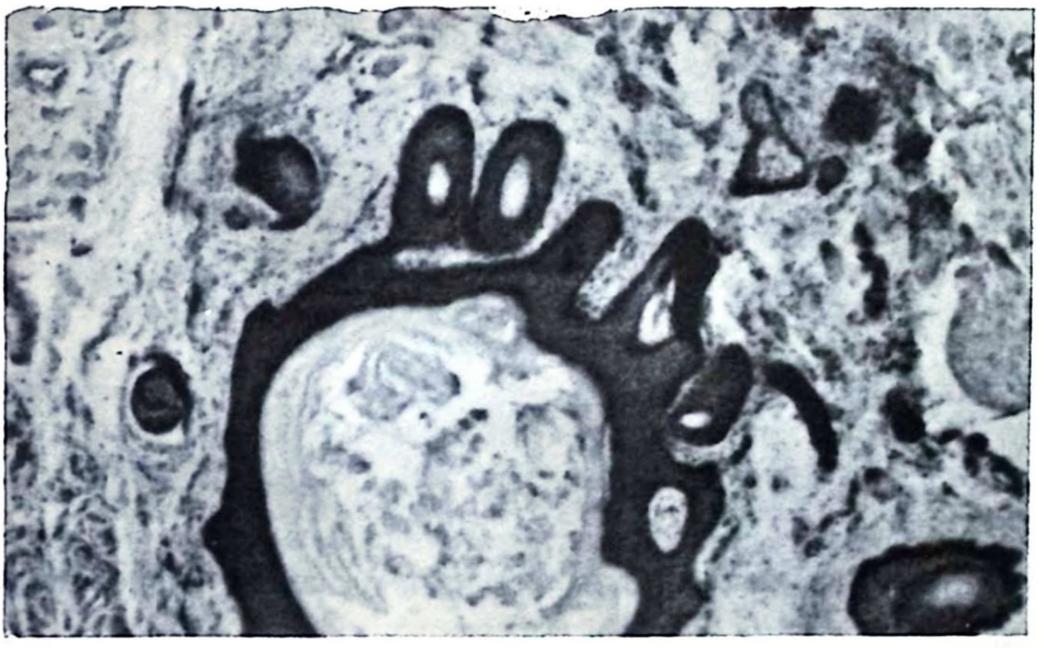


Fig. 2: Histologic section of the dermal tract revealed stratified squamous epithelium in papillary structure with underlying sebaceous glands, hair follicles, capillaries, and keratinocious lamellas inside. H.E X 40.

Case 2

A 13-year-old girl was referred in September 1993 with mild back pain increasing during exercise and with the complaint of serous discharge occurring occasionally in the lumbosacral area. Neurologic examination was normal. The skin 2 cm to the right of the midline was dark bule and included a dermal sinus. Plain radiographs demonstrated posterior fusion defects in the sacral vertebrae, lamina and spinous process anomaly at L5, and a wide interpeduncular space at the L3-5 levels.

Omnipaque myelography disclosed a large caudal sac and filling defect on the posterior surface of the dural sac at the L4-5 disc level. CT delineated a hyperdense mass located intrathecally. Myelo CT demonstrated the attachment of a dermal sinus tract with the mass at L4-5 (Fig. 3) and conus visualization at the L3-4 disc level.

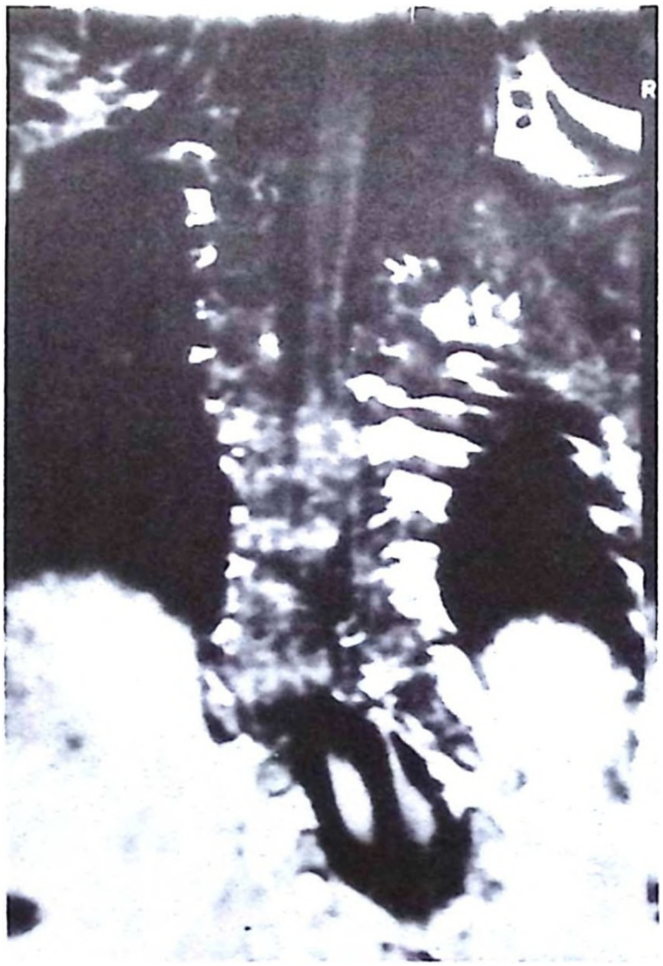


Fig. 3: Myelo CT demonstrates the attachment of dermal sinus tract with the mass at the L5 lavel.

Complete laminectomy was performed on the L4 and L5 vertebrae and partially on the L3. A dermal sinus tract extending obliquely on the right side perforated the L5 lamina and dura and adhered to a tumor in the subarachnoid space. Upon opening the dura, a conus at the L3-4 disc level was revealed. With microsurgery the sinus tract and the mass with a separate peduncle that attached the conus were removed completely. The filum terminale, which was thick and short and ended at the posterior dural surface at the L5 level, was sectioned. The patient's postoperative course was uneventful, and after three months she had no complaint.

Microscopic sections of the tumor showed that the cyst was lined by stratified squamous epithelium, and underlying it connective tissue, fat and muscle tissue, vascular structures, sympathetic ganglia and lymphoid tissue were observed. Inside the cyst there were keratinocious lamelles (Fig. 4).

Fig. 4: Histologic section of the tumor showed connective tissue, muscle and fat tissue and peripheral nerves H.E X 40.

Discussion

Congenital malformations appearing in association with occult spinal dysraphism may occur as separate entities or in a more complex pattern. Various combinations of these malformations have been reported in the literature[2, 6, 11-15].

The manifestations associated with these malformations are caused by traction, compression, myelodysplasia or rarely inflammation[3]. The clinical presentation of a patient with occult spinal dysraphism differs according to the pathological events involved. It can manifest itself with a neuromusculoskeletal syndrome[6, 16], sphincter disturbance[2, 4, 8], spinal curvature[6, 16], cutaneous malformation[2] or back pain[8, 17]. In most cases the neurological impairment of the lower extremities or of bladder function is superimposed on the clinical situation.

The tumor observed in the thoracic region of the first case had two cystic components and a tract between them. The cystic component of the tumor and the structure of the tract included this tumor in the category of dermoids. Although the spinal cord typically reunites below the cleft in most patients with diastematomyelia, extension as split cords through the conus is extremely rare[3, 16]. A second form of occult spinal dysraphism including low conus and dermal sinus in the lumbosacral area and hydromyelia in the thoracic region is rarely seen[3, 14].

Another unusual occurrence is dextrocardia, which is usually observed in a patient with complete situs inversus or with Kartagener's syndrome. When it occurs without situs inversus, associated malformations are the rule[18]. Dextrocardia in association with occult spinal dysraphism was thought to be an extremely unusual finding in our patient, as no report was found related to this association in the literature.

Fifty percent of patients reported in the literature with a dermal sinus had an inclusion tumor. These tumors were classified as epidermoid in 13 percent, dermoid in 83 percent and teratoma in four percent of cases[3]. Teratoma are tumors having various cellular or organized components of normal derivatives from more than one germ layer[19]. Occasionally it has appeared in the spinal canal as a separate case[20], and occurrence with occult spinal dysraphism is extremely rare[21, 22]. The tumor found in the second patient was included in the category of benign teratomas due to its composition, which had components arising from the mesoderm and ectoderm. Congenital spinal dermal sinuses above the sacrococcygeal area are quite rare, and almost all reported cases have occurred along the dorsal midline[23].

Plain roentgenograms are abnormal in the majority of patients[1, 2, 16]. MR imaging is simplifying the investigation of occult spinal dysraphism to a great degree, and it was possible to achieve an accurate diagnosis in most cases[2, 9, 10]. Some difficulty was observed with the MRI scan in differentiating the exact level o the conus from a thickened filum and in solving the diagnostic problem of ruling out a retethered conus[10]. Despite these pitfalls, the MRI scan has become the first investigative technique in evaluation of occult spinal dysraphism[9, 24]. When it is not available or there is high suspicion about the pathology despite normal MRI findings, the patient should be further investigated with water-soluble contrast myelography and Myelo CT[3, 11, 15].

Any child with a cutaneous stigmata, foot deformity, spinal curvature or failure to gain sphincter control at an appropriate age should undergo radiologic investigation regardless of neurologic status. If the initial deficit is due to true myelodysplasia, no improvement can be achieved surgically, but secondary effects due to traction or compression may be prevented or reversed[2, 6, 15, 16]. Thus, early diagnosis and surgical therapy to prevent future or progressive neuronal deficit are mandatory.

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