Isolated flat capillary midline lumbosacral hemangiomas as indicators of occult spinal dysraphism

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Object. Historically, cutaneous stigmata representative of occult spinal dysraphism (OSD) have included lumbar hemangiomas. Frequently, this skin change is found in conjunction with other cutaneous alterations such as dermal sinus tracts and subcutaneous lipomas. Debate has recently surrounded the question of whether these skin changes in isolation might indicate underlying spinal disease. The authors reviewed their experience in their most recent 120 cases in which OSD was diagnosed.

Methods. The authors retrospectively reviewed records obtained in 120 patients with OSD. They found that many of the patients reviewed harbored only a flat capillary hemangioma as an indicator of OSD. In 21 patients (17.5%) with only midline lumbosacral flat capillary hemangiomas, underlying OSD was present. No single variety of OSD had a higher incidence of association with this single cutaneous stigma.

Conclusions. Based on their experience, the authors recommend magnetic resonance (MR) imaging evaluation in cases involving this skin lesion in isolation to discern the potential for surgically significant spinal cord anomalies. Prospective studies are now needed to examine MR images obtained in all children with this lesion in the midline lumbosacral spine and assess for OSD.

Key Words • hemangioma • capillary • lumbosacral spine • spinal dysraphism

Spinal dysraphism refers to a distinct group of congenital anomalies characterized by a failure of midline structures of ecto- and mesodermal origin to fuse. Spina bifida aperta, which is obvious at birth, is contrasted with spina bifida occulta, which is covered with skin and frequently associated with cutaneous stigmata over the midline lumbosacral spine.

Cutaneous hemangiomas occur in approximately 2.6 to 12% of newborns. This broad range certainly represents the generic appellation of the term “hemangioma” to a variety of vascular anomalies such as macular stains and telangiectasias. These blemishes are thought to represent angioblastic tissue that fails to unite formally with the developing vasculature of the body. Lumbar hemangiomas have historically been listed as cutaneous stigmata indicative of underlying OSD much as facial port-wine stains are indicators of leptomeningeal angiomatosis in Sturge–Weber syndrome. These cutaneous signatures, commonly known as nevus flammeus neonatorum when indicating underlying spinal dysraphism, are often found in association with other skin changes such as subcutaneous lipomas. The authors of a recent study, however, concluded that isolated lumbosacral “strawberry nevi” do not appear to indicate underlying dysraphic states. We review our experience with isolated lumbosacral flat capillary hemangiomas and determine the incidence of associated lumbosacral OSD.

Clinical Material and Methods

We retrospectively reviewed our records obtained in the most recently treated 120 patients in whom lumbosacral OSD was diagnosed (Children’s Hospital, Birmingham, AL, and Pediatric Neurosurgery, Madison, WI). Patients were observed to have isolated midline flat capillary hemangiomas of the lumbosacral spine as the only cutaneous manifestation of their underlying OSD. We performed the Student t-test to determine if any single form of spinal dysraphism such as lipomyelomeningocele or SCM had a greater association with these isolated midline flat capillary hemangiomas.

Results

In a review of patient’s charts, we found that isolated lumbosacral flat capillary hemangiomas over the midline were documented in 21 patients. These port-wine stains were not velvety, were irregularly outlined, and blanched with pressure. There were nine male and 12 female pa-
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Table 1 provides a summary of all forms of OSD found in association with these skin markings in our series. Figures 1 to 4 are imaging and photographic examples of the various isolated flat capillary hemangiomas and their underlying intradural disease observed in this series. No single form of spinal dysraphism was associated with isolated flat capillary hemangiomas of the lumbosacral spine (p > 0.05).

Discussion

Occult spinal dysraphism is often associated with superficial stigmata of the skin. The six most common stigmata include focal hirsutism, capillary hemangioma, dermal sinus, subcutaneous lipoma, rudimentary appendage, and atretic meningocele. The occurrence of skin stigmata is not surprising considering the pluripotent nature of the early developing ectoderm.

Cutaneous angiomas ordinarily found in other parts of the body are unusual in the lumbosacral region, where they have been associated with OSD but are often seen in conjunction with other skin findings, such as focal hirsutism and subcutaneous lipomas. Capillary hemangiomas are thought to be the least sensitive indicator of intradural disease, with approximately a 10% incidence of associated intradural anomalies in the lumbar region. Some authors have criticized the use of “capillary” and “cavernous” to describe hemangiomas because these lesions arise from the same cell type. Serna, et al., conducted a histological examination of a capillary angioma obtained from the lumbar region in a patient with a lumbar dermal appendage and subcutaneous lipoma. This examination revealed dilated vessels with a regular disposition in the capillary dermis surrounded by a marked fibrosis and a significant mast cell infiltrate. They concluded that this corresponded to the involutorial phase of a capillary angioma. Allen, et al., and Selden, et al., have recently stated that isolated lumbosacral strawberry nevi do not indicate the presence of OSD. In their study, 20 consecutive newborns with lumbosacral strawberry nevi underwent ultrasonography and/or MR imaging. In those with isolated nevi, there were no MR imaging findings indicative of OSD. The authors concluded that these nevi alone may not indicate underlying dysraphism of the spine. More than half of the patients, however, did not undergo MR imaging of the caudal spine to confirm the negative ultrasonographic findings despite the fact that in two patients MR imaging revealed lesions that the other modality did not. In fact, Hughes, et al., performed ultrasonography of the spine in 85 infants. Of these, 15 patients (mean age 40 days) underwent follow-up MR imaging. Ultrasonography failed to demonstrate four of four posterior dermal sinuses, three of four fatty terminal filae, one of one terminal lipoma, two of four partial sacral ageneses, and three of four syringohydromyelia. Agreement between ultrasonography and MR imaging was good (~90%), however, for the detection of low-lying spinal cords.

Albright, et al., evaluated seven children with lumbar cutaneous hemangiomas (six true raised “hemangiomas” [ectatic vessels in the papillary dermis] and one flat capillary hemangioma). In all patients tethered spinal cords were observed intraoperatively and in two children there were associated cutaneous stigmata of OSD. Whether the one child with a flat capillary hemangioma had this cutaneous stigma in isolation is unclear. Confusion with dermatological terms is certainly an issue when these lesions are described. Morelli categorized vascular birthmarks into hemangiomas, of which a superficial variety is the “strawberry,” and malformations, of which capillary lesions (port-wine stains) are found.

Ben-Amitai, et al., reviewed data in 28 of 3623 neonates in whom sacral nevus flammeus simplex was diagnosed. Ultrasonography of the lumbosacral area performed in 25 patients (89%) revealed SCM in one. In conclusion, they recommended that neonates with this sin-

**Fig. 1.** Four examples of isolated lumbosacral flat capillary hemangiomas in our series.
gle cutaneous finding undergo examination for OSD because this may constitute the only cutaneous sign of spinal dysraphism.

Although isolated lumbosacral flat capillary hemangiomas are occasionally found in patients with normal intradural anatomy, our analysis of the retrospective data suggests that a large number will harbor surgically significant OSD. Interestingly, in our cohort, syringohydromyelia was present in eight cases, three of which were terminal syringes. Because the majority of patients evaluated in early childhood with surgically significant forms of OSD will be neurologically normal and frequently the first manifestation of neurological compromise is the development of an irreversible neurogenic bladder, prospective intervention is warranted.9,10 Hoffman, et al.,6 reported that deterioration occurred in 85% of their pediatric patients with untreated lipomyelomeningocele by the age of 5 years. Similarly, Guthkelch5 studied 35 children and at long-term follow-up examination found that in more than 90% neurological function had been lost by the age of 15 years. Moreover, the unusual nature of a heman-

Fig. 2. Sagittal T₂-weighted MR image of the thoracolumbosacral region obtained in the patient in Fig. 1 center left.

Fig. 3. Axial T₁-weighted MR image of the fatty terminal filum demonstrated in a patient with an isolated flat capillary hemangioma.

Fig. 4. Intraoperative photograph obtained in a patient with an isolated flat capillary hemangioma, demonstrating one of the meningocele manqué (arrow) that tethered the spinal cord posteriorly.
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gioma located in this region should prompt the clinician to undertake further investigation. One must also distinguish hemangiomas from mongolian spots, which are pigmented blue or black and are most commonly found in the sacral region of Asian newborns.11

Conclusions

Based on our experience, midline lumbosacral hemangiomas should undergo MR imaging to discern the potential for surgically significant spinal cord anomalies. Prospective studies are now needed to evaluate MR imaging studies in all children with this lesion in the midline lumbosacral spine and to determine the incidence of OSD.

References

5. Guthkelch AN: Diastematomyelia with median septum. Brain 97:729–742, 1974

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