Neonatal Lumbosacral Ulceration Masking Lumbosacral and Intraspinal Hemangiomas Associated with Occult Spinal Dysraphism

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A 12-day old full-term neonate was evaluated for a lumbosacral ulceration that had developed 4 days previously. Per parental report, an oval pink patch with a dimple, surrounded by a tuft of hair, was noted overlying the midline lumbosacral spine at birth. An ultrasonography (US) scan performed at an outside hospital on day 2 of life showed no evidence of underlying spinal dysraphism. On examination, there was a 4-cm by 1.5-cm oval deep ulceration over the midline back extending from the lumbar to the sacral spine. There were several bright-red papules noted along the inferior border of the ulceration (**Figure**, A).

Because of the location of the lesion over the midline lumbosacral region and previous history of a dimple with a tuft of hair, magnetic resonance imaging (MRI) of the spine was performed that demonstrated a low-lying conus medullaris terminating at the L4 level. A T2 hypointense thickened filum terminale was noted below the conus medullaris and adhered to the posterior aspect of the thecal sac diagnostic of a tethered spinal cord (**Figure**, B). There was a small, blind ending sacral dermal sinus extending from within the ulceration to the base of the spinal cord. No intradural masses were evident.

Due to concerns of delayed wound healing of the ulceration and fecal contamination of the dermal sinus leading to possible meningitis, a diverting colostomy was performed. A pediatric dermatology consultation was obtained. Consultants concluded that, given the history of the previous red plaque, the association with a tethered spinal cord, and the red papules along the inferior rim of the ulceration, the diagnosis was ulcerated infantile hemangioma (IH) with occult spinal dysraphism (OSD). The findings of a full neurologic assessment and kidney US scan were normal.

Oral propranolol given at a dose of 0.94 mg/kg/day led to rapid healing of ulceration and resolution of vascular papules within 6 weeks of initiating treatment. As the result of observed improvement in the size of the IH, the parents discontinued propranolol. Tethered cord release surgery was performed at 12 weeks of age. At 16 weeks, the infant presented with a new 2-cm by 3-cm red plaque in the right perianal area (**Figure**, C). There was no ulceration or pain. Topical timolol maleate 0.5% gel-forming solution 2 drops of twice daily was initiated to attempt to decrease the size of the IH.

The cutaneous IH regressed after 8 weeks of timolol. Repeat postoperative MRI was performed at 6 months of age and revealed a large intraspinal IH, including a small nodular intrathecal lesion at the lower end of conus medullaris and a large extradural IH in the sacral spinal canal with extraspinal extension into presacral and gluteal regions (**Figure**, D). Oral propranolol at 2 mg/kg/day was started and timolol discontinued. Colostomy was reversed at 8 months of age. After 6 months of treatment with oral propranolol, MRI at 12 months of age showed marked decrease in the size of the intraspinal IH. Oral propranolol was discontinued at 15 months of age, and repeat MRI at 18 months of age demonstrated continued decrease in volume of the intraspinal IH. Our patient is currently 19 months old, and he continues to develop normally without neurologic deficit. Repeat MRI is planned at 24 months of age to monitor for spinal cord retethering.

Overview of IH

Cutaneous vascular lesions may occur at or shortly after birth. The 2014 International Society for the Study of Vascular Anomalies[1] classified vascular anomalies into 2 groups: the proliferative vascular tumors and nonproliferative vascular malformations.[2] IHs are the most common type of proliferative vascular tumor, occurring in 1%-5% of neonates.[3] Risk factors include female sex, preterm birth, and low birth weight.[4] At birth, IHs often are absent or can be present as a faint, erythematous, telangiectatic patch. Rapid proliferation of IH occurs between 5.5 and 7.5 weeks of age followed by slower growth between 6 and 9 months of age, after which the IH slowly involutes over years.[5]

Less than 10% of IHs occur in the lumbosacral or perineal region, where ulceration is more common.[6] Ulceration is the most common complication in IHs, occurring in up to 15.8% of infants, especially during periods of rapid hemangioma proliferation.[7] Pain is the most significant morbidity of ulcerated IHs. The infants are inconsolable, often with impaired feeding and disrupted sleep. Rarely, IHs can manifest as a

IH	Infantile hemangioma
MRI	Magnetic resonance imaging
OSD	Occult spinal dysraphism
US	Ultrasonography

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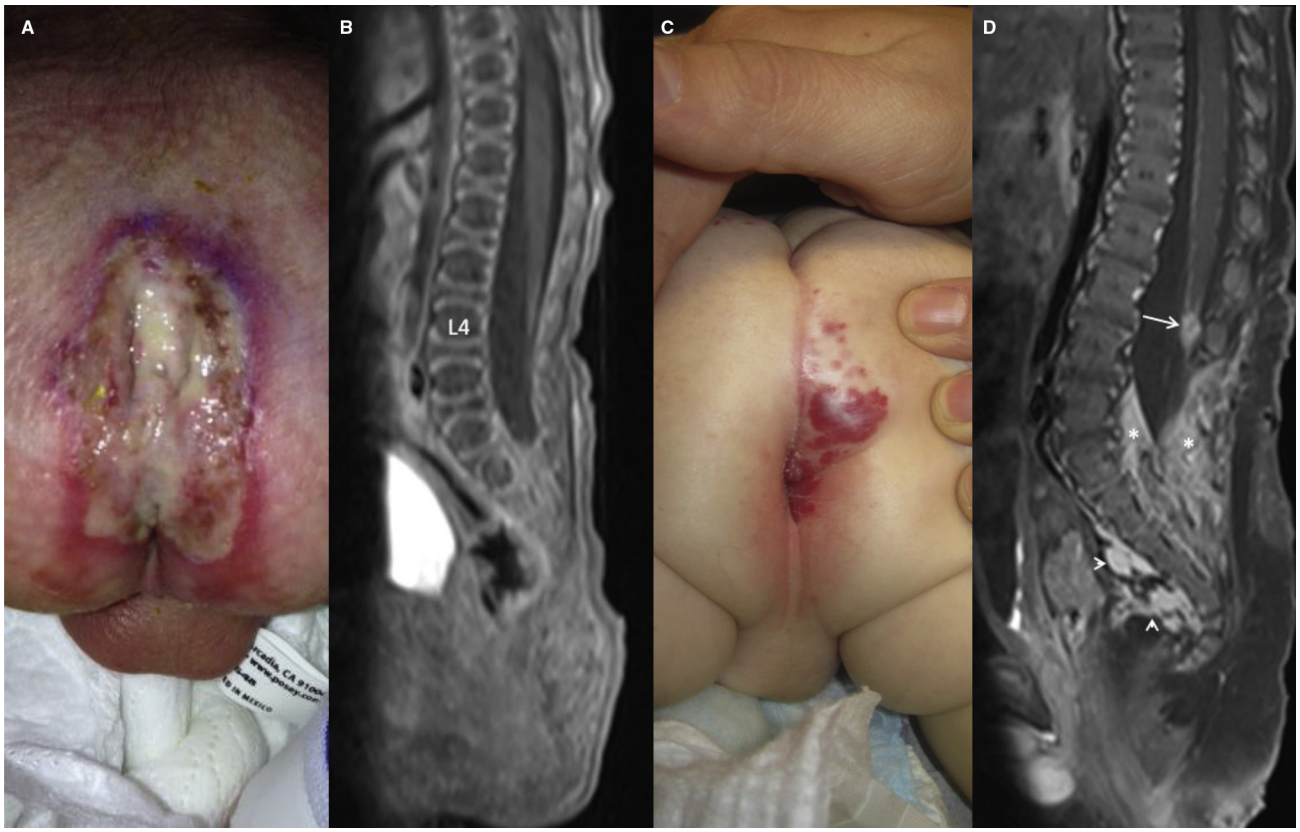

Figure. **A,** Lumbosacral ulcerative plaque with surrounding red vascular rim was noted on initial examination. **B,** Midline sagittal contrast-enhanced, T1-weighted, fat-saturated image of the lumbosacral spine at presentation reveals low-lying conus at L4 vertebral level suggestive of tethered cord. **C,** Recurrence of lumbosacral hemangioma after discontinuation of oral propranolol. **D,** Midline sagittal contrast-enhanced, T1-weighted, fat-saturated image of the lumbosacral spine at 6 months of age shows new nodular enhancing lesion at the lower end of the conus (*arrow*) compatible with intrathecal hemangioma. In addition, there is a large hemangioma in the epidural space in sacral spinal canal (*asterisks*) with presacral extension (*arrowheads*).

congenital or neonatal ulceration in the lumbosacral and perineal regions and diagnostic features of hemangioma may be absent.[8] Careful inspection of surrounding skin for classic red vascular papules of IH or telangiectases may be a clue to underlying diagnosis.

Treatment of IH

Although most IHs do not require treatment because they regress spontaneously, IHs that are ulcerated, impair vital functions (periorbital, laryngotracheal, intraspinal), or have the potential to cause permanent disfigurement (nasal-tip IHs) require treatment. Ulcerated IHs are challenging to treat. A multifaceted approach with effective topical wound care, local pain control, and prevention of wound infection is required. Oral propranolol also has been shown to be rapidly effective in the treatment of ulcerated IHs.[9] For recalcitrant ulcerations, some experts have reported success with becalpermin[10] (topical platelet-derived growth factor) gel, pulse-dye laser,[11] and topical timolol.[12] Bacterial colonization of the wound can delay wound healing. In our patient, a diverting colostomy was performed to prevent further wound contamination, promote wound healing, and potentially reduce risk for meningitis as there was a known dermal sinus ending in the area of ulceration.

Oral propranolol hydrochloride is approved by the United States Food and Drug Administration for the treatment of IHs and usually is continued until 10-16 months of age depending on the response.[13,14] Rebound growth of the IH is more common with discontinuation before 12 months of age (6%-19%) compared with after 12 months of age.[15] Rebound of the cutaneous IH in our patient was noted only 10 days after discontinuation of propranolol. Superficial IH (less than 2 mm thick) may be treated effectively with off-label use of topical timolol maleate.[16] The degree of systemic exposure from topical application through neonatal skin is unknown, and absorption may be enhanced when applied to ulcerated IH. Adverse events attributable to propranolol and timolol are rare and include bronchospasm, bradycardia, hypotension, and hypoglycemia.[17]