Takayasu arteritis coexisting with scalp necrosis, alopecia, and sterile osteomyelitis

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ABSTRACT

A twelve-year-old girl with classical features of Takayasu arteritis presented with scalp ulceration and osteomyelitis. Her computed tomography (CT) of the head revealed an extensive ulcerated lesion over the left high parietal region with lytic destruction of the outer and inner tables of the skull. Because of full-thickness calvarial bone involvement, chronic osteomyelitis, and ulcerated scalp lesion, she underwent debridement of involved bone along with the margin of normal skin. During surgery, underlying dura was found to be not involved, and a transposition flap was done for reconstruction. Histopathology did not reveal any evidence of bacterial infection or granulomas. Sterile osteomyelitis of the skull associated with alopecia and scalp necrosis has not been reported with typical Takayasu disease. Family physicians should be vigilant to keep this as a differential diagnosis in nonhealing osteomyelitis, not responding to antibiotics, or showing any evidence of infection.

Keywords: Scalp ulceration, sterile osteomyelitis of skull, Takayasu arteritis

Introduction

In India, Takayasu arteritis is frequently encountered in young females, a rare form of large vessel vasculitis affecting the aorta and its branches.¹² Sterile osteomyelitis rarely occurs in autoimmune diseases. We describe a rare case of Takayasu arteritis associated with sterile osteomyelitis of skull bone with scalp ulceration or necrosis.

Case Report

A twelve-year-old girl with classical features of Takayasu arteritis presented with scalp ulceration over the left parietal region with lytic destruction of tissues [Figure 1] and osteomyelitis. She had a history of sudden onset left upper limb monoparesis ten months back, her magnetic resonance imaging MRI of brain was suggestive of acute infarcts in the right frontal, parietal, temporal, occipital region, and basal ganglia, with significant thickening and narrowing of the right brachiocephalic trunk, right subclavian, right common carotid arteries, left common carotid [Figure 2] and left subclavian artery and was treated with steroids. On admission, the patient had absent radial pulsations, diminished femoral pulsations, and an ulcer over the left parietal region of the scalp with necrotic tissues [Figure 1]. Her computed tomography (CT) of the head revealed an extensive ulcerated lesion over the left high parietal region with lytic destruction of the outer and inner tables of the skull. CT aortography was done, which revealed a normal arch of the aorta, brachiocephalic trunk, and bilateral subclavian arteries showed circumferential thickening causing 60-70% luminal stenosis, left common carotid artery showed narrowing of 60-70% from its origin, the right common carotid artery showed 80% luminal narrowing, right vertebralbasilar artery showed 60-70% luminal narrowing, left subclavian artery distal to left vertebral artery showed luminal narrowing, right subclavian artery showed narrowing over its origin, descending aorta and lower limb of sudden onset left upper limb monoparesis ten months back, her magnetic resonance imaging MRI of brain was suggestive of acute infarcts in the right frontal, parietal, temporal, occipital region, and basal ganglia, with significant thickening and narrowing of the right brachiocephalic trunk, right subclavian, right common carotid arteries, left common carotid [Figure 2] and left subclavian artery and was treated with steroids. On admission, the patient had absent radial pulsations, diminished femoral pulsations, and an ulcer over the left parietal region of the scalp with necrotic tissues [Figure 1]. Her computed tomography (CT) of the head revealed an extensive ulcerated lesion over the left high parietal region with lytic destruction of the outer and inner tables of the skull. CT aortography was done, which revealed a normal arch of the aorta, brachiocephalic trunk, and bilateral subclavian arteries showed circumferential thickening causing 60-70% luminal stenosis, left common carotid artery showed narrowing of 60-70% from its origin, the right common carotid artery showed 80% luminal narrowing, right vertebralbasilar artery showed 60-70% luminal narrowing, left subclavian artery distal to left vertebral artery showed luminal narrowing, right subclavian artery showed narrowing over its origin, descending aorta and lower limb...
arteries were normal in caliber. There was no evidence of infection, but ESR and CRP were high.

Because of full-thickness bone involvement, chronic osteomyelitis, and ulcerated scalp lesion, she underwent debridement of involved bone along with the margin of normal skin; during surgery, underlying dura was found to be not involved and transposition flap was done for reconstruction. Surgical biopsies from the margins and base of the ulcer and surrounding skull bones were taken, and intraoperative findings were suggestive of extensive destruction of the skull was present, with no underlying dural involvement was seen. On histopathological examination of the debrided bone moderately dense lymphoplasmacytic infiltrate, tuoton type giant cells and few necrotic debris were seen [Figure 3a]. Surrounding areas showed congested vessels and stromal edema [Figure 3b]. There was no evidence of bacterial infection or granuloma.

Discussion

Sterile osteomyelitis is an important differential diagnosis to be kept in autoimmune inflammatory diseases and osteomyelitis cases in the pediatric age group. It can resemble infectious osteomyelitis radiologically and histopathologically. Chronic recurrent multifocal osteomyelitis (CRMO) is an inflammatory disorder that primarily affects children. The clinical presentation is an insidious onset of bone pain with or without fever. Laboratory studies typically reveal nonspecific evidence of inflammation. Family physicians and primary care providers must be aware of CRMO as a diagnostic entity when evaluating a child who presents with clinical and histologic evidence of osteomyelitis. There is often a diagnostic delay and unnecessarily prolonged treatment with antibiotics. About 25% of individuals with CRMO have an associated inflammatory disorder like palmar-plantar pustulosis, psoriasis vulgaris, inflammatory bowel disease, pyoderma gangrenosum, inflammatory arthritis, Takayasu arteritis.[1-3] Scalp skin lesions and sterile osteomyelitis of the skull associated with Takayasu arteritis have not been reported. The exact cause is not definitely known. Bone lesions were correlated with the areas where high arteritis activity.[3] Ischemic hypoxia due to arterial stenosis may be responsible for the skeletal abnormality.[3] However, the locations of the bone lesions in other cases, including ours, were not related to the artery lesions, suggesting that osteomyelitis was caused by active inflammation rather than hypoxia. In cases with no definitive evidence of infection or if the presumed infection is not responding, sterile osteomyelitis should be considered in the pediatric age group with Takayasu arteritis.

Key points

• Scalp necrosis or osteomyelitis of skull bone may be a manifestation of vasculitis syndrome per se if secondary infection is ruled out.
• Early treatment of skull bone osteomyelitis is important to prevent breach of inner table of bone and infection spreading to duramater and brain.

A presumed chronic osteomyelitis associated with Takayasu disease and not responding to antibiotics might be a case of sterile osteomyelitis.
Patient consent and approvals
Patient written informed consent and permission taken for publishing case report.

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Conflicts of interest
There are no conflicts of interest.

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