CASE REPORT

Childhood Sarcoidosis Presenting as Recurrent Facial Palsy

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Background: Recurrent facial palsy in a patient merits investigation for underlying etiology. **Case characteristics**: 8-year-old boy with erythematous itchy skin lesion and recurrent facial palsy. **Observation**: He had a past history of aseptic meningitis and nephrocalcinosis. Raised angiotensin converting enzyme levels, interstitial lung disease on CT chest, and non caseating granulomas on skin biopsy clinched the diagnosis of sarcoidosis. **Message**: Multisystem involvement and recurrent lower motor facial nerve palsy is a clinical clue for sarcoidosis.

Keywords: *Bell's palsy, Etiology, Multisystem involvement*.

Sarcoidosis in children is often missed because of its myriad presentations and rarity. We present a child with sarcoidosis who had recurrent facial palsy and multisystem involvement.

CASE REPORT

An 8-year-old boy presented with itchy, erythematous plaque like skin lesions involving the buttocks and extremities for 8 months. He was treated with topical steroids without significant improvement. He had a past history of various illnesses starting from 3 years age. At 3 years, he had a left sided lower motor facial palsy which resolved on oral steroids. He had 3 further episodes of facial palsy at 4 years, 4.5 years and 6 years treated each time with short courses of oral steroids. At 4 years of age he had a prolonged febrile episode for 1.5 months associated with headache, vomiting and facial palsy. Investigations at that time showed an ESR of 71, CRP 20 (normal <6 mg/L), SGPT - 7 U/L, ANA negative, HIV negative. Mantoux was positive, CXR showed bilateral infiltrates. His MRI brain was normal. CSF examination had revealed 142 cells/cc, 90 lymphocytes, sugar 52 mg/dL, and protein 65 mg/dL. CSF PCR for tuberculosis was negative. Family history for tuberculosis was positive. He was treated with antitubercular therapy for 1 year with initial 6 weeks of oral steroids and made a good recovery. One month after starting anti-tubercular therapy he developed leaking of clear fluid from the sacral region. MRI spine revealed a hypointense track at S1-2 level extending from the thecal sac up to the skin suggestive of dorsal dermal sinus. The sinus tract was excised. Histopathology of the surgical specimen showed fibrocollagenous tissue and granulation tissue with lymphocytic infiltrates but no granulomas.

At presentation to us at 8 years, besides skin lesions and a 3 cm hepatomegaly he had no other significant findings. His blood pressure was normal and eye examination including slit lamp for uveitis was normal. Additional history of effort intolerance, significant hair loss and non-specific bony pains over the last 6 months were elicited on directed questioning.

A skin biopsy revealed lichenoid granulomatous infiltrates with small and medium sized granulomas located close to the epidermis and in perifollicular location. These granulomas had epitheloids cells, giant cells, surrounding lymphocytes and no caseation. A diagnosis of granulomatous dermatitis either sarcoidosis or lichen scrofulosum was considered.

Chest *X*-ray and a Mantoux test were normal. Complete blood count and SGPT was normal limits. ESR was 46 mm FHR. Angiotensin converting enzyme (ACE) levels was elevated 119U/L (normal 7-52). High resolution computed tomography (HRCT) of the chest showed confluent areas of ground glass opacities in bilateral lung parenchyma with mild diffuse interlobar interstitial septal thickening noted bilaterally. Tiny calcifications were noted in the aortopulmonary region with few enlarged bilateral hilar nodes measuring 1-1.2 cm, suggestive of sarcoidosis. Serum creatinine, calcium and 25-hydroxy vitamin D levels were normal.

A diagnosis of sarcoidosis was made and he was started on oral steroids and hydroxychloroquine. His skin lesions, dyspnea on exertion and non-specific body pains disappeared in a month and scalp hair became more luxurious. His steroids are being slowly tapered and he

remains asymptomatic during follow over the past 6 months. His ACE levels are now normal.

DISCUSSION

This child presented with a long history of over five years with recurrent facial nerve palsy, aseptic meningitis, nephrocalcinosis and skin lesions. A recent review reported the incidence of sarcoidosis in children as 0.22–0.27 per 100,000 children/year [1]. From India isolated case reports and short case series of pediatric sarcoidosis have been reported [2]. Sarcoidosis in children is probably missed due to its indolent course and presentation to different medical subspecialties for varying symptoms.

Clinical symptoms in sarcoidosis include fever, weight loss, cough, arthritis, and erythema nodosum. In children, the disease frequently involves the lungs, lymph nodes, eyes, skin, liver and spleen [3]. In a study, in patients with sarcoidosis, 48% had neurological symptoms and peripheral facial nerve palsy was the commonest abnormality [4]. Renal involvement in sarcoidosis includes nephrocalcinosis and interstitial nephritis.

Recurrent Bells palsy has been reported in 2-9% of all cases. Other causes of recurrent facial palsy are underlying tumour, Behchets syndrome, Lyme disease, hypertension, Ramsay Hunt syndrome, Melkersson-Rosenthal syndrome, or familial occurrence [5].

The diagnosis of sarcoidosis is based on a compatible clinical and/or radiological picture; histological evidence of noncaseating granulomas and exclusion of other diseases. Close differentials in this child include early onset sarcoidosis and Blau syndrome, which are monogenic autoinflammatory syndromes. However, the classic triad of arthritis, dermatitis and uveitis was incomplete in this patient. Other differentials include granulomatosis with polyangiitis. However, it is a disease of older age group, the renal involvement is usually glomerulonephritis and not nephrocalcinosis, and ACE levels are not elevated in this condition. However, HRCT chest may show characteristic findings [6]. Many diagnostic modalities are available once sarcoidosis is suspected, including bronchoalveolar levage and Fibreoptic bronchoscopy [7]. Serum ACE is useful in monitoring the course of disease.

In case of steroid toxicity or steroid resistant disease, immunosuppresive agents like methotrexate and hydroxychloroquine have been used with some success [8]. A monoclonal antibody against TNF alpha, infliximab, and a combination of infliximab and mycophenolate mofetil have been recently used to treat those resistant to steroids [9]. Multiorgan and CNS involvement is associated with a poor prognosis while facial nerve palsy has been reported to carry a good prognosis [10]. In conclusion, sarcoidosis must be considered in a child with recurrent facial nerve palsy especially in the setting of multi-organ involvement.

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REFERENCES

1. Hoffman AL, Milman N, Byg KE. Childhood sarcoidosis in Denmark 1979-1994: Incidence, clinical features and laboratory results at presentation in 48 children. Acta Paediatr. 2004;93:30-6.
2. Gupta SK. Pediatric sarcoidosis in India. Indian J Pediatr. 2001;68:931-5.
3. Shetty AK, Gedalia A. Childhood sarcoidosis: A rare but fascinating disorder. Pediatr Rheumatol. 2008;6:16-25.
4. Stern BJ, Krumholz A, Johns C, Scott P, Nissim J. Sarcoidosis and its neurological manifestation. Arch Neurol. 1985;42:909-17.
5. Salman MS. Facial paralysis and Bell's palsy in children: Current diagnostic concepts and management controversies. Primary Psychiatry. 2003;10:67-70.
6. Ors F, Gumus S, Aydogan M, Sari S, Verim S, Deniz O. HRCT findings of pulmonary sarcoidosis; relation to pulmonary function tests. Multidiscip Respir Med. 2013;8:8.
7. Lynch JP 3rd, Ma YL, Koss MN, White ES. Pulmonary sarcoidosis. Semin Respir Crit Care Med. 2007;28:53-74.
8. Cimaz R, Ansell BM. Sarcoidosis in the pediatric age. Clin Exp Rheumatol. 2002; 20:231-7.
9. Moravan M, Segal BM. Treatment of CNS sarcoidosis with infliximab and mycophenolate mofetil. Neurology. 2009; 72:337-40.
10. Delaney P. Neurologic manifestations in sarcoidosis: Review of the literature, with a report of 23 cases. Ann Intern Med. 1977; 87:336-45.