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CASE REPORT

Hyperimmunoglobulin E syndrome with juvenile dermatomyositis and calcinosis

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Abstract Juvenile dermatomyositis (JDM) is a rare childhood disease with autoimmune association. Environmental factors are known to trigger JDM in genetically susceptible individuals (Schmieder et al., Dermatol Online 6:3, 2009). Calcinosis is a well-established complication of JDM. Prevalence is higher in children (30–70%; Özkaya et al., Erciyes Med J 30(1):40–43, 2008). Hyperimmunoglobulin E syndrome is a primary immunodeficiency syndrome with multiple recurrent abscess formation and raised serum immunoglobulin E levels. We report a case of JDM with calcinosis cutis universalis with hyperimmunoglobulin E syndrome. With a previous similar case report (Min et al., Korean J Intern Med 14:95–98, 1999), this could well be a new sequence syndrome where abscesses are the trigger for the onset of JDM.

Keywords Calcinosis cutis universalis · Hyperimmunoglobulin E syndrome · Juvenile dermatomyositis · Multiple abscesses

Summary

Juvenile dermatomyositis (JDM) is a rare autoimmune vasculopathy of childhood with probable underlying genetic susceptibility. Environmental triggers have been reported [1]. Definitive diagnostic criteria are based on those established by Bohan and Peter in 1975 [2]. Calcinosis as a complication of JDM is seen in 25–50% of patients with an even higher prevalence in children (30–70%) [3]. The hyperimmunoglobulin E syndrome is a rare primary immunodeficiency syndrome characterized by recurrent abscesses and elevated levels of serum IgE. Abscess formation can trigger development of dermatomyositis. Hyperimmunoglobulin E syndrome, as a cause of recurrent abscesses in a case with juvenile dermatomyositis, has been reported once previously [4]. We report another case of juvenile dermatomyositis with calcinosis cutis universalis associated with hyperimmunoglobulin E syndrome. The similarity to the previous report suggests that JDM associated with hyperimmunoglobulin E may not have been coincidental, and this could represent a new sequence syndrome.

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Case report

The 11-year-old Hindu male was the third child born to non-consanguineous parents. His problems began to be noticed at around 3.5 years of age. He had low grade intermittent fever with generalized myalgia and fatigue along with occurrence of multiple abscesses all over the body. Abscesses were recurrent and involved mostly the trunk and extremities. His weakness progressed gradually with difficulty in climbing stairs and frequent falls. He became completely bed ridden by 6 years of age. Recurrent abscess formation continued with suppuration. He had empyema thoracis twice requiring drainage of right pleural cavity at 5 and 8 years of age.

At 11 years of age, he remained bright and alert though he had severely wasted muscles of the extremities and flexion contractures at elbows and knees. His hips and knees are flexed to the chest. There were multiple abscesses with suppuration all over the body, multiple-matted cervical

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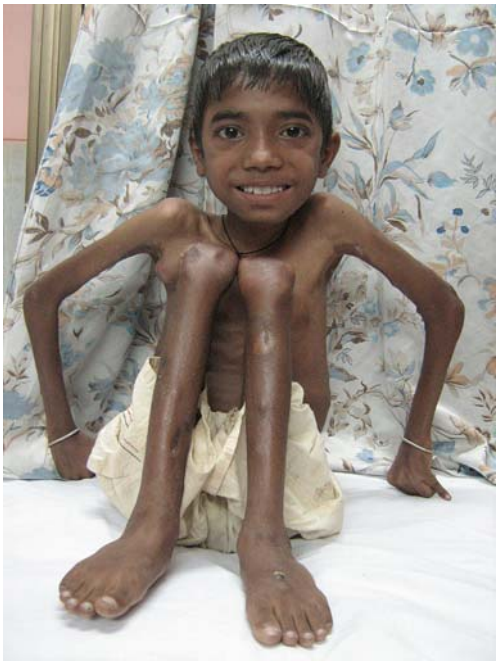

Fig. 1 Photograph of our patient with JDM with hyperimmunoglobulin E syndrome showing contractures at elbows and knees with nodular calcifications and abscesses in the whole body

Table 2 Laboratory investigations (microbiology)

Parameters	Results
Sputum for AFB, 3 samples	Negative
Urine routine microscopy	Normal
Urine culture and sensitivity	Sterile
Blood culture	Sterile
Pus culture	Sterile
CD4	757 cells/μl
CD3	1,241 cells/μl
CD4/CD3 ratio	0.61
NBT	Normal
HIV	Non-reactive

lymph nodes (axillas, neck) and multiple irregular nodular bony hard swelling all over the body (extremities, axillas, chest, abdomen; largest being 8×4 cm; Fig. 1).

His weight was 21.5 kg against an expected weight for his age of 36 kg (50th percentile). Respiratory system, cardiovascular system, and abdomen were otherwise normal. Higher mental functions were normal, as were all the cranial nerves. Sensory system was normal. Upper limb power was 2–3/5 in both limbs. Lower limb power could not be assessed because of contractures. Reflexes could not be elicited.

Tables 1 and 2 show the list of laboratory investigations done in the child along with their normal reference values.

Table 1 Laboratory investigations (hematology, biochemistry, and immunoassay)

Parameter	Patient values	Reference value
Hemoglobin (g/dl)	9	11.5–18
Hematocrit	30.2	35–54
TLC (mm^{-3})	10,100	4,500–10,500
DLC	P67/L30/M01/E02	P40-75/L20-50/M2-10/E1-6
Platelet count (mm^{-3})	448,000	150,000–450,000
PBS study	Normocytic normochromic anemia	
Serum bilirubin (mg/dl)	0.6	0.2–1.0
SGPT (U)	27	5–35
Alkaline phosphatase (U)	118	100–280
Serum albumin (g/dl)	4.3	3.7–4.9
Serum calcium (mg/dl)	10	8.5–10.5
Serum phosphorus (mg/dl)	4.8	2.5–5.5
Serum PTH (intact; pg/ml)	21.9	10–69
Serum uric acid (mg/dl)	2.60	4–7.5
Blood urea (mg/dl)	23	20-40
Serum creatinine (mg/dl)	0.6	0.2–1.5
Serum LDH (U)	833	225–450
Serum creatine kinase (U)	326	25–190
Serum IgG (mg/dl)	3,316.4	570–1,410
Serum IgA (mg/dl)	143.3	65–260
Serum IgM (mg/dl)	176.9	60–175
Serum IgE (IU/ml)	1,739.1	<100

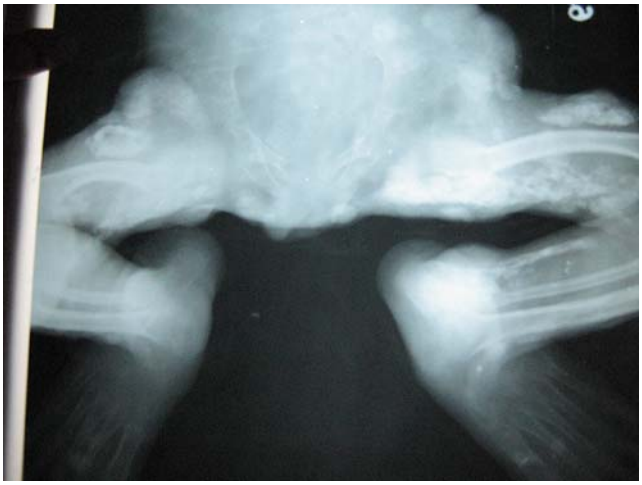

Fig. 2 X-ray of trunk and lower extremities showing multiple diffuse calcifications

Serum lactate dehydrogenase and creatine kinase levels were elevated. Serum IgE level was markedly raised (1,739 IU/ml). Serum IgG was also raised (3,316 mg/dl).

Radiology showed multiple diffuse subcutaneous, intramuscular, and lymph node calcification, involving many parts of the body (Fig. 2). Computed tomography scan of chest and abdomen also showed multiple calcific density foci in subcutaneous, intramuscular, and lymph node areas with muscle atrophy. FNAC right lower thoracic region swelling was suggestive of inflammatory lesion. Biopsy showed extensive dermal calcification (calcinosis cutis) and myositis, with intramuscular calcium deposition. Nerve conduction velocity study findings were suggestive of muscular dystrophy or polymyositis.

Discussion

Diagnostic criteria for JDM are based on those established by Bohan and Peter in 1975, which include a characteristic skin eruption, symmetrical proximal muscle weakness, elevated muscle enzymes, pathological muscle histology, and myopathic electromyographic changes [2]. The presence of three of these criteria characterizes definite JDM [1]. Calcinosis is a known complication of JDM. Calcinosis occurs in 30% of patients with JDM. The serum calcium and phosphorus levels in such children are normal, and the calcinosis is dystrophic in nature. The sites most frequently affected are the elbows, knees, digits, and extremities, although it may occur virtually anywhere over the body. The onset of calcinosis is most often 1–3 years after illness onset but has been reported to occur from the time of illness, and sometimes, onset may be delayed by up to 20 years. There are four subtypes of dystrophic calcification described in the literature in association with JDM [5], and our patient had calcinosis cutis universalis—a classic presentation with involvement of all big joints as well as diffused skin and muscular plane involvement.

Though JDM has a probable underlying genetic susceptibility, environmental factors such as bacterial and viral infections as well as exposure to ultraviolet light are important trigger factors for the disease onset [1]. So, probably hyperimmunoglobulin E recurrent infection has acted as a trigger for the onset of vasculopathy associated with JDM in our patient. The raised IgG levels would also have been due to repeated infections.

Our case of JDM associated with hyperimmunoglobulin E syndrome with recurrent abscess formation, seen along with the previous case reported with the same problems [4], suggests that this could be a previously unrecognized sequence syndrome.

Disclosures None

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