CASE REPORT

Cutaneous polyarteritis nodosa in a child and a review of the literature

H Mocan1, MC Mocan2, H Peru1 and Y Özoran3

Departments of Paediatrics1 and Pathology3, Faculty of Medicine, Karadeniz (Black Sea) Technical University, Trabzon, and Hacettepe University Hospital2, Ankara, Turkey

The cutaneous form of polyarteritis nodosa in children is extremely rare. Findings are usually limited to the skin, muscles and joints. It has a benign but often chronic course. We describe an 8-y-old girl with cutaneous PAN, with extensive livedo reticularis on lower and upper extremities, tender subcutaneous nodules, arthralgia and right ankle swelling. Skin biopsy revealed vasculitis of small and medium-sized blood vessels characterized by fibrinoid necrosis. The use of prednisolone resulted in clinical improvement initially, but recurrence occurred during tapering. She showed marked improvement with additional high dose methyl prednisolone monthly.

Polyarteritis nodosa (PAN) is a necrotizing vasculitis involving small and medium-sized arteries, that most often affects the kidneys, heart and liver, but can affect any organ system (1). In the cutaneous form of PAN (CPAN), tender subcutaneous nodules with high fever, arthralgias and myalgias occur without major organ system involvement (1, 2). The histopathological futures are those of a nodular arteritis, involving medium-sized arteries in the deep reticular dermis, showing extensive fibrinoid necrosis and vascular destruction.

Case report

An 8-y-old girl was admitted to paediatric unit with mottled red appearance on her extremities, fever, right ankle swelling and arthralgia. The cutaneous lesions had appeared continuously during the previous 3 y, with milder exacerbations and additional leg pain that was not responsive to analgesics. Two days before admission, she developed slight right ankle swelling and fever. The patient’s history was negative for abdominal pain, change in bowel habits, urine colour or hypertension. Her medical and family histories were unremarkable.

On physical examination, she was febrile and had an extensive livedo reticularis with patchy maculopapular, erythematous eruption on the lower and upper extremities and trunk. There were several 0.5–2 cm tender nodules on feet, ankles and distal legs (Fig. 1). There was neither organomegaly nor hypertension. The right ankle was slightly swollen and tender.

Laboratory data included a haematocrit of 29%, white blood cell count 8700/μl with 64% neutrophils, 28% lymphocytes and 8% monocytes; platelet count 498 × 109/l, normal electrolyte and creatinine levels, normal urinalysis, and erythrocyte sedimentation rate (ESR) of 38 mm/h. Hepatitis B surface antigen, antinuclear antibody, rheumatoid factor, anticardiolipin antibody, C3 and C4 complement components, liver function tests, electrocardiogram, chest roentgenogram, throat and urine cultures were either normal or negative. Antistreptolysin O (ASO) titre was 1:400 Todd units. ANCA was not tested. A skin biopsy showed inflammatory cells primarily polymorphonuclear leukocytes infiltrating deep dermal, muscular arteries and fibrinoid necrosis (Fig. 2). Treatment was started with prednisolone 2 mg/kg/d for 2 months and tapered over 2 months. She improved initially, but recurrence occurred during tapering. Following subsequent treatment with high-dose prednisolone, the painful nodules and joint pain again showed only minimal improvement. High-dose methyl prednisolone (HDM) (30 mg/kg) orally was given monthly. She showed marked improvement and has remained asymptomatic during the past 3 months, except for livedo reticularis.

Discussion

Borrie (1) and Diaz-Perez (2) described a clinically and histopathologically distinct variant of PAN that they called benign CPAN, which in children is extremely rare (3–13). The first case was reported by Verbov (3) in 1980. It is a rare form of vasculitis that appears to be limited primarily to the skin, muscles and joints. In contrast to the systemic form of the disease (14) it is characterized by the absence of visceral lesions and a relapsing but benign course. Typical skin findings are painful red nodules, especially on the lower extremities though other lesions, including ulcers,
gangrene, urticaria, livedo reticularis or bullae, resemble those of systemic PAN. With onset in the first decade of life, a higher incidence of peripheral gangrene has been reported (12). The diagnosis of CPAN is made clinically and biopsy examination of affected skin by demonstration of fibrinoid necrosis of the small and medium arteries similar to our case (Table 1).

Laboratory tests have usually been unremarkable, except for elevated ESRs (10). Guillet et al. (6) reported that while an elevated ESR in systemic PAN is a bad prognostic sign, it indicates a recurrent but benign course in cases of CPAN. The aetiology of CPAN is unknown, but the presumption of immunocomplex mediation in some is based on the demonstration of IgM and C3 deposits in lesional biopsy specimens (2) and the detection of circulating immune complexes (15). Serum complements have usually been normal (7). Associations with streptococcal infections also suggest immunocomplex mediation (2, 3, 13). However, this possible association was not noted in a large series of 31 patients with systemic PAN (14). Although an association with hepatitis B surface antigen has been reported in adults with CPAN (7), this association in childhood CPAN has not been described. Crohn’s disease and chronic ulcerative colitis have both associated with CPAN (7, 15).

Possible association of benign CPAN in children was noted with diphtheria–pertussis–tetanus immunization, drugs, wasp sting and falciparum malaria (12). The unique coexistence of CPAN plus antiphospholipid antibodies and p-ANCA was also reported in a child (11). Aspirin (3, 6), prednisolone (5, 7, 9) and methotrexate (8), alone or in combination, have relieved acute exacerbations, except for failure of aspirin therapy in the case reported by Jones et al. (4). Many adult patients have appeared to respond to NSAIDs (2). Remissions may also suggest immunocomplex mediation (2, 3, 13). However, this possible association was not noted in a large series of 31 patients with systemic PAN (14). Although an association with hepatitis B surface antigen has been reported in adults with CPAN (7), this association in childhood CPAN has not been described. Crohn’s disease and chronic ulcerative colitis have both associated with CPAN (7, 15).

### Table 1. Childhood cutaneous polyarteritis nodosa: literature summary.

<table>
<thead>
<tr>
<th>Author</th>
<th>No. of cases</th>
<th>Age (y)</th>
<th>Sex</th>
<th>Skin lesions</th>
<th>Other findings</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Verbov (3) [1980]</td>
<td>1</td>
<td>5.5</td>
<td>F</td>
<td>Nodules, maculopapular rash, livedo reticularis</td>
<td>Facial oedema, polyarthralgia, joint swelling</td>
<td>Aspirin</td>
</tr>
<tr>
<td>Jones (4) [1985]</td>
<td>5</td>
<td>F</td>
<td>Nodules</td>
<td>Swollen PIP*</td>
<td>Aspirin (failed), prednisone</td>
<td></td>
</tr>
<tr>
<td>Volk (5) [1986]</td>
<td>1</td>
<td>9</td>
<td>F</td>
<td>Nodules</td>
<td>Swollen knees, ulcerative colitis</td>
<td>Prednisone</td>
</tr>
<tr>
<td>Guillet (6) [1987]</td>
<td>1</td>
<td>5</td>
<td>F</td>
<td>None initially; nodules</td>
<td>Shoulder, elbows</td>
<td>Aspirin</td>
</tr>
<tr>
<td>Moreland (7) [1990]</td>
<td>1</td>
<td>15</td>
<td>F</td>
<td>Nodules, gangrene</td>
<td>Elbow, wrist, knee swelling</td>
<td>NSAID, prednisone</td>
</tr>
<tr>
<td>Jorizzo (8) [1991]</td>
<td>1</td>
<td>12</td>
<td>F</td>
<td>Nodules, livedo reticularis</td>
<td>?</td>
<td>Methotrexate</td>
</tr>
<tr>
<td>Magilvay (9) [1977]</td>
<td>1</td>
<td>13</td>
<td>M</td>
<td>Nodules, rash</td>
<td>Arthralgias, myalgias</td>
<td>Prednisone</td>
</tr>
<tr>
<td>Sibbery (10) [1994]</td>
<td>2</td>
<td>6, 12</td>
<td>2F</td>
<td>Nodules (2)</td>
<td>Nodules (2) papular rash (1)</td>
<td>Pain, swelling in knees (1) Aspirin (failed) (1) Arthralgias, polyarthritits (1) Prednisone (2)</td>
</tr>
<tr>
<td>Sheth (13) [1994]</td>
<td>4</td>
<td>2.5–14</td>
<td>3F/1M</td>
<td>Nodules (4) Cyanotic digits/livedo reticularis (2)</td>
<td>Arthralgia/arthritis (4) myalgia/myositis (4)</td>
<td>Aspirin (4), failed (2) Prednisone (3) IVGG (1), failed Penicillin (4)</td>
</tr>
<tr>
<td>Kumar (12) [1995]</td>
<td>10</td>
<td>1.25–10</td>
<td>3F/7M</td>
<td>Nodules, ulcerations, vesiculobuluous lesions (10), gangrene (8), livedo reticular (4)</td>
<td>Arthralgia/arthritis (7)</td>
<td>Prednisone (7) Prednisone + cytotoxic drugs (3)</td>
</tr>
<tr>
<td>Present case</td>
<td>1</td>
<td>8</td>
<td>F</td>
<td>Nodules, livedo reticularis, maculopapular rash</td>
<td>Arthralgia, ankle swelling</td>
<td>Aspirin (failed) Prednisone, HDMP**</td>
</tr>
</tbody>
</table>

+ Not reported as cutaneous polyarteritis nodosa.
* PIP, indicates proximal interphalangeal joint; NSAID, nonsteroidal anti-inflammatory drug.
** High dose methyl prednisolone.
occur spontaneously and recurrences are common (2). Our case also did not respond to aspirin before admission. We observed clinical improvement with prednisolone for 2 months, but she had recurrence during tapering. She showed marked improvement in joint pain and skin lesions with HDMP (30 mg/kg/monthly) in addition to prednisolone treatment and had no recurrences during the past 4 months. We suggest HDMP should be considered if prolonged corticosteroid therapy is required for interspersed clinical course with remissions and exacerbations.

From the literature review, we would recommend that the diagnosis of CPAN should be considered in a child with fever, tender subcutaneous nodules, typical skin findings and arthralgia/arthritis. We recommend that they are specific enough so the clinicians do not have to make extensive investigations for systemic involvement, i.e. renal biopsy, UCG, ANCA analysis etc. Because of the benign course of CPAN and the known side effects of the drugs used to treat systemic PAN, the recognition of CPAN is important and over-treatment should be avoided.

References
4. Jones SK, Lane AT, Golitz LE, Weston WL. Cutaneous periarteritis nodosa in a child. AJDC 1985; 139: 920–2

Received May 30, 1997. Accepted in revised form Sep. 24, 1997