Severe abdominal involvement as the initial manifestation of cutaneous polyarteritis nodosa in a young girl

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ABSTRACT

We report a young girl who developed ingravescent intestinal symptoms as the first manifestation of cutaneous polyarteritis nodosa (PAN) while the typical skin nodules developed later during the disease course. Cutaneous PAN predominantly affects children and presents with crops of painful skin nodules in the medial aspect of the foot, often preceded by sore throat. Visceral manifestations including gut involvement are commonly associated with the classical form of PAN while they are rarely reported in the cutaneous form. In our patient the severity of the abdominal symptoms required a laparoscopy, which revealed diffuse erythematous swelling of the intestine on the serosal side. The administration of penicillin and steroids was followed by a dramatic improvement in the disease course. Chronic anterior uveitis developed 4 months after the disease onset and responded to local treatment. At a 2-year follow-up the girl is in good condition under prophylaxis with benzathine-penicillin with no recurrence of the illness.

Our case confirms that cutaneous PAN is often related to streptococcal infection, and suggests that ASO titers should be determined in children with vasculitides to ensure a timely diagnosis and treatment of the condition if present.

Introduction

Cutaneous polyarteritis nodosa (PAN) is a necrotizing vasculitis affecting the medium-sized and small blood vessels. The clinical manifestations of classical polyarteritis nodosa are fever, arthralgia, peripheral neuropathy, myalgia and visceral involvement (gastrointestinal tract, kidney, heart, and central nervous system). The term “cutaneous PAN” is reserved for the cutaneous form of the disease, which is characterised by typical skin alterations and mild systemic symptoms. The histology of the cutaneous form is identical to that of systemic PAN. Cutaneous PAN predominantly affects children and presents with crops of painful skin nodules and livedo reticularis, often preceded by sore throat or ear infection (1,2). The crops of nodular rash are particularly common on the medial side of the foot, but may occur at any site. Transient arthritis affecting the knees and ankles is not uncommon. Most cases run a benign course, but relapses can also occur.

We report the case of a young girl affected with cutaneous PAN whose first clinical manifestation was a rapidly ingravescent involvement of the gastrointestinal tract, while the cutaneous alterations developed later during the disease course.

Case report

A 6-year-old girl was referred to our Paediatric Clinic in June 1998, with a one-month history of high fever, abdominal pain and general malaise. On admission she was pale and severely ill. Physical examination revealed a diffuse erythema of the oropharynx and a diffuse tenderness of the abdomen, without liver or spleen enlargement. Blood tests showed: erythrocyte sedimentation rate (ESR, Westergren) 108 mm/h, WBC 7,700/mm³ with neutrophils 78.9%, and a C-reactive protein of 9.56 mg/dL (nv < 0.5 mg/dL). Blood, stool, and urine cultures were negative, as were serological tests for viral and bacterial infections. A broad spectrum intravenous antibiotic empiric treatment was introduced, without any improvement. Fever reached 40°C, and the girl developed bloody diarrhoea. Abdominal CT and ultrasound scans and small bowel barium follow-through were unremarkable, while rectal endoscopy showed multiple shallow ulcers on the rectal mucosa. Histology showed the presence of an aspecific inflammatory infiltrate and excluded inflammatory bowel disease. Since the abdomen became intractable an explorative laparoscopy was performed, which revealed diffuse erythematous and a swollen gut on the serosal side.

The patient was given total parenteral nutrition with no amelioration. Intravenous methylprednisolone was then administered, which was followed by a dramatic improvement in the clinical findings and laboratory tests. Steroids
were progressively tapered, with a rapid recurrence of fever and arthralgia. A bone marrow biopsy showed no evidence of immature or abnormal cells. In July 1999 the patient developed arthritis of both ankles and crops of painful erythematous nodules on the medial aspect of both feet (Fig. 1). Blood tests showed again an elevation of inflammatory parameters (ESR 90 mm/h) and low haemoglobin levels (7.8 g/dL). All other laboratory results were within normal limits, including hepatitis C screening, liver function tests, ANA, ANCA, and urinalysis. Blood pressure was normal. A renal ultrasound did not show any abnormalities; a renogram was not performed. A skin biopsy showed evidence of leukocytoclastic vasculitis (Fig. 2). Cutaneous PAN related to streptococcal infection was suspected, and the ASO titer was found to be raised to 1330 U. A throat culture was positive for Group A Streptococcus. Benzathine-penicilllin (1,200,000 U) and oral steroids (prednisolone 1 mg/Kg/day) were administered, with a dramatic improvement in the clinical findings. The girl was discharged on low dose steroids, and penicillin prophylaxis was started (Benzathine-penicilllin 1,200,000 U every 3 weeks). Later in the disease course she developed headache and dyspnoea. Magnetic resonance imaging (MRI) and MR angiography excluded a vasculitic involvement of the cerebral vessels, while a slit lamp examination showed inflammation in the anterior chamber of both eyes which responded to a brief course of local steroids. The patient is currently not taking prednisone, and is still in full remission.

Discussion
Our case is of interest because severely ingravescent gut involvement was the inaugural manifestation of cutaneous PAN while the typical skin painful nodules became evident only after 5 weeks. The severity of the abdominal symptoms required a laparoscopy, which revealed a diffuse erythematous swelling of the intestine on the serosal side. A barium follow-through and an intestinal biopsy were not consistent with the diagnosis of inflammatory bowel disease. Broad-spectrum antibiotic treatment was unable to control the disease activity. The appearance of crops of skin painful nodules on the medial aspect of the foot associated with swelling of both ankles raised the suspicion of cutaneous polyarteritis and and there fore we looked for streptococcal infection. A significant increase in ASO titers since the onset of abdominal symptoms and the development of skin nodules raised the suspicion of cutaneous polyarteritis and and therefore we looked for streptococcal infection. A significant increase in ASO titers since the onset of abdominal symptoms and the development of skin nodules confirmed the link between vasculitis and Group A streptococcal infection. The administration of penicillin and steroids was followed by a dramatic improvement in the disease course. Chronic anterior uveitis developed 4 months after the disease onset and responded to local treatment with mydriatic and steroid drops. After two years of follow-up the girl is in good condition under prophylaxis with benzathine-penicilllin and with no recurrence of the disease. The possible relationship between childhood PAN and group A streptococcal infection has already been proposed by Fink (3, 4) and Blau (5). Fink first reported 7 children affected with PAN, 6 of whom had a preceding sore throat or otitis and 5 of whom showed elevated ASO titers. In the English literature (6) 12 children with Group A streptococcus-related vasculitis are reported. The mean interval from disease onset to diagnosis was 1 to 12 weeks, and in all patients there was a history of preceding sore throat. In more than 80% of the patients non-specific abdominal pain was reported, and in one patient uveitis. In our patient the benign course of the disease following treatment with penicillin and low dose steroids, together with the absence of renal, cardiac and neurologic involvement confirm the diagnosis of the cutaneous form of PAN: in the classical form of PAN visceral involvement is predominant, and in the majority of patients the disease course is severe despite aggressive treatment with immunosuppressive drugs (7).
Our case confirms the notion (8) that cutaneous PAN is often related to streptococcal infection, and suggests that ASO titers should be determined in children with vasculitides in order to diagnose the condition and introduce appropriate antibiotic treatment. In our patient the first clinical manifestation of cutaneous PAN was involvement of the gastrointestinal tract. The presence of shallow ulcerations of the colonic mucosa with aspecific inflammatory changes was documented. To the best of our knowledge this finding at disease presentation has never been reported before in the literature.

References