www.jpnim.com Open Access eISSN: 2281-0692
Journal of Pediatric and Neonatal Individualized Medicine 2014;3(2):e030202
doi: 10.7363/030202
Published online: 2014 Jun 14

Answer

Polyarteritis nodosa in a 21-month-old child: answer

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Keywords

Quiz, polyarteritis nodosa, child, kidney.

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How to cite

Sanna A, Gerosa C, Ambu R, Puddu M, Nurchi AM, Faa G. Polyarteritis nodosa in a 21-month-old child: answer. J Pediatr Neonat Individual Med. 2014;3(2):e030202. doi: 10.7363/030202.

Answers

1. Multiple renal infarcts.
2. Fibro-obliterative arteritis suggestive for polyarteritis nodosa.

Introduction

Childhood polyarteritis nodosa (PAN) is a necrotizing vasculitis, affecting small and medium size blood vessels. This condition was first described by Kussmaul and Maier in 1866 [1]. Although comparatively rare in childhood, it is the most common form of systemic vasculitis in children [2, 3]. PAN includes two different subtypes, the classical systemic form presenting with a wide range of clinical manifestations including dermatologic, musculoskeletal, nervous, renal, and gastrointestinal systems and the more frequent cutaneous form (CPAN) that involves only the skin [4]. The main clinical features of PAN are malaise, fever, weight loss, skin rash, myalgia, abdominal pain and arthropathy [5, 6]. Systemic involvement is variable, but the skin, the musculoskeletal system, the kidneys and the gastrointestinal tract are most prominently affected, with cardiac, neurological and respiratory manifestations occurring less frequently [7]. However, clinical manifestations can be very confusing, with absence of conclusive diagnostic evidence in the early phase and sometimes in the late phase of the illness [8]. The etiology of PAN remains unclear, but there are data to support roles for hepatitis B [9] and reports of a higher frequency of exposure to parvovirus B19 and cytomegalovirus in PAN patients compared with control populations [10]. However, in childhood, associations between PAN and these infections or other conditions are rare. Evidence has emerged suggesting that bacterial superantigens may play a role in some cases [11]. Here we report the clinico-pathological findings of a 21-month-old child affected with PAN, with particular emphasis on the severity of renal pathological lesions.

Pathological findings

At macroscopy, the kidney showed an irregular surface with depressed zones red in color, intermingled with pale gray areas, highly suggestive for kidney infarcts. The histological study of kidney sections at low power showed multiple ischemic infarcts in the deep cortex. The ischemic infarcts had ill-defined borders and were different in size: small and large infarcts were observed (**Fig. 1**). At higher power, severe changes were observed in interlobular arteries. The main lesion was an occlusive endarteritis that caused the obstruction of the lumen (**Fig. 2**). Fibrinoid necrosis and infiltration of lymphocytes and polymorphonucleates were also observed in the wall of small-medium sized arteries (**Figures 3** and **4**).

Discussion

The case here reported shows some peculiar findings that deserve some consideration.

1. The early insurgence (21 months) induces to consider this case exceptional, the median age of patients with PAN being 9.5, ranging from 5 up to 14 [12].
2. A severe kidney damage characterized our child, contrasting with the rarity of renal failure in classic PAN [12].
3. No drug assumption was present in the history of our patient, contrasting with recent reports on the association between minocycline therapy and renal PAN insurgence [13].

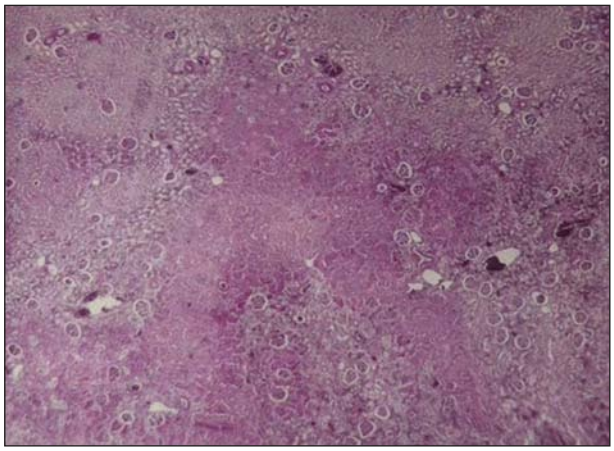

Figure 1. Ischemic infarct of the renal cortex: panoramic view. Weigert-Van Gieson, x 100.

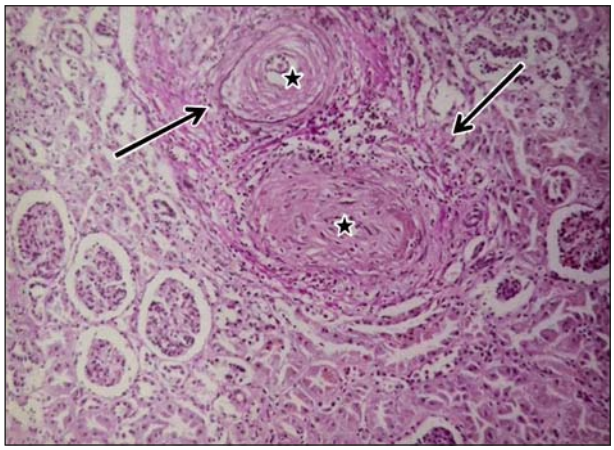

Figure 2. Renal interlobular arteries (arrows) showing severe architectural changes characterized by a limphocytic infiltrate in the adventitia and by a fibro-obliterative intimal proliferation with a marked reduction of the arterial lumen (stars). Weigert-Van Gieson, x 100.

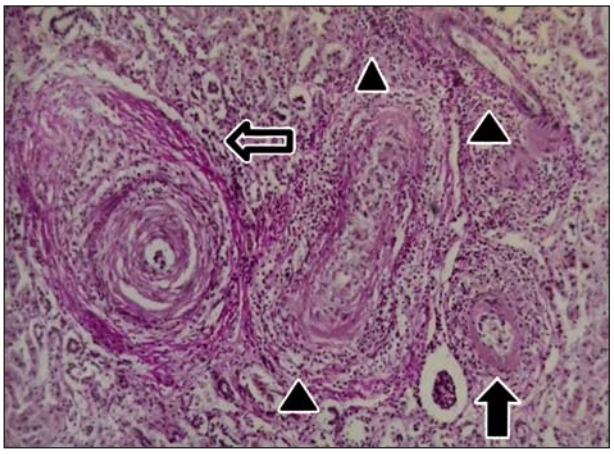

Figure 3. Active obliterative arteritis of small/medium size renal arterioles. Note the different stage of the process in adjacent vessels: on the right, active necroinflammation with fibrinoid necrosis (arrow); on the left, fibro-obliterative arteritis (open arrow): in the center, an occlusive inflammatory reaction, leading to complete occlusion of the arterial lumen (arrowheads). Weigert-Van Gieson, x 100.

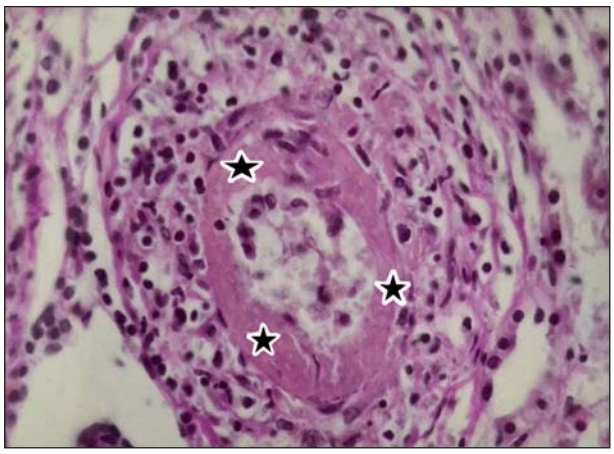

Figure 4. At high power, the complete disarrangement of the entire wall of an affected arterial vessel is better shown. Stars: fibrinoid necrosis. Weigert-Van Gieson, x 100.

Declaration of interest

The Authors declare that there is no conflict of interest.

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