Reviewer’s report

Title: A case of Polyarteritis Nodosa limited to the right calf muscles, fascia and skin

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Reviewer: JOSE HERNANDEZ-RODRIGUEZ

Which of the following best describes what type of case report this is?: Unexpected or unusual presentations of a disease

Has the case been reported coherently?: Yes

Is the case report authentic?: Yes

Is the case report ethical?: Yes

Is there any missing information that you think must be added before publication?: Yes

Is this case worth reporting?: Yes

Is the case report persuasive?: No

Does the case report have explanatory value?: Yes

Does the case report have diagnostic value?: Yes

Will the case report make a difference to clinical practice?: Yes

Is the anonymity of the patient protected?: Yes

Comments to authors:

The authors reported a patient with vasculitis of the right calf muscle and its surrounding fascia and skin who was successfully treated with local methylprednisolone injections and oral azathioprine.

This case resembles to those described in the literature as “isolated vasculitis of calf muscles”. In patients with this condition, cutaneous lesions, including purpura and erythema, over the inflamed calf muscle have been reported. Steroids are initially effective in all cases, but during their tapering local relapses frequently occur. For relapsing patients cytotoxic agents are usually required. Thus, long term follow-up is essential to confirm the limited extent of this vasculitis.

Several major aspects have to be considered:
1. The name of Polyarteritis nodosa (PAN) has to be used for any systemic vasculitis satisfying the ACR classification and/or Chapel Hill nomenclature criteria for PAN. Histological features of vasculitis and/or angiographic changes typical of medium sized vessel vasculitis in an appropriate setting of patients, in whom a multiorgan vasculitis is suspected, may be diagnostic of PAN. Although in the literature, there is a remarkable number of articles talking about “limited or localized PAN”, the correct term for these cases would be “isolated or limited necrotizing (or the predominant histological change) vasculitis of a specific organ or territory”. In the current case, the use of “limited form of PAN” should be avoided along the text, and instead, “Limited or localized necrotizing vasculitis of the right calf muscle and fascia” is recommended.

2. Because the skin was not proved to be affected by vasculitis, the authors must admit, at least, that the cutaneous changes in this patient could also be secondary to tissue remodeling and edema due to the subjacent inflammatory process. The authors must consider that if vasculitis had been found in more than one organ-system, such as the skin and muscle, this would have led to the diagnosis of systemic vasculitis.

3. Because the patient initially received glucocorticoids and azathioprine (intralesional Depo-Medrone has well known systemic effects), was being still treated with azathioprine at the end of the follow-up, which was as short as 5 months, the systemic involvement cannot be entirely ruled out and the limited nature of the process should be, at least, questioned. Although I think the case resembles vasculitis limited to the calf muscle, please include this limitation.

4. Page 4. The sentence: “Histopathological features were consistent with polyarteritis nodosa” is not appropriate. The previous description was “In one fragment of muscle there was small vessel vasculitis with fibrinoid changes in the vessel wall and intense perivascular and focal mural chronic inflammatory changes” and looking at the figure 2, indeed, necrotizing vasculitis involves a small vessel. However, by definition, PAN is a vasculitis that affects medium sized vessels (without affecting smaller vessels – according to Chapel Hill definition). As a result, histologically, this case would correspond to a necrotizing small vessel vasculitis (ANCA associated, cryoglobulinemic…all negative… so, unclassifiable vasculitis??). Therefore, I would name and describe this case as “necrotizing (small vessel) vasculitis of the right calf muscle and fascia”.

5. The abstract and conclusion should be modified accordingly.

Minor points
1. The four Depo-Medrone courses were administered every 1,2 or 4 weeks? This should be stated, as well as the dose of azathioprine given to the patient.
2. Please add chest-X ray results
3. Page 4: When describing MRI changes, imaging descriptions and MRI sequences used, and what these changes were suggestive of should be placed instead of talking about “extensive abnormalities” or “localize abnormality”. Please do the same in the figure legend of figure 3 (in this figure legend, MRI of the right leg should be also replaced by “MRI of both legs” since both legs appear
in the figure).

4. Because it is not completely sure that MRI signal enhancement of the affected muscles have to correspond to edema (vasculitis, which is the main lesion, leads to a direct inflammation that can be confounded with edema), in the last sentence edema could be replaced by inflammatory changes.

5. In Figure legend 2, please add “... in the right calf muscle.”

Quality of written English: Acceptable

Declaration of competing interests:

I declare that I have no competing interests