

firmatory.<sup>1</sup> In these cases the clinical symptoms are mild and most of the patients reach adult life. Unfortunately, the functional obstruction, dilation and hypertrophy gradually increase proximal to the lesion. The dilated bowel is no longer able to propel the faeces distally. The patient has severe constipation or, as in our case, acute intestinal obstruction. The latter condition has been reported in only 1 patient in a large series from the Mayo Clinic and only a few other cases.<sup>1</sup>

The number of cases with local aganglionosis is uncertain but we believe that there is a group of patients classified in the subgroup of 'megacolon unassociated with aganglionosis', and other unclassified cases of megacolon that could be considered in this group, after other acquired conditions have been ruled out. The criteria for diagnosis in this group of patients are clinicopathological. A long history, especially in a man, of chronic constipation and abdominal distension with, on examination, a normal rectum, barium enema and sigmoidoscopy is combined with the histological finding in any part of the colon of a localized segment with no ganglion cells; the sigmoid appears more frequently affected<sup>5</sup> and there are normal ganglia in the rectal biopsy specimen. Every effort must be made to confirm the diagnosis, and treatment must be limited to

resection of the diseased segment of colon, with primary or delayed anastomosis.<sup>3</sup>

We conclude that investigation of all cases of unexplained chronic constipation and abdominal distension in an adult should include the possibility of Hirschsprung's disease, even in cases where the rectal biopsy is negative.

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# Widespread superficial thrombophlebitis as a manifestation of secondary syphilis — a new sign

## A report of 2 cases

H. F. JORDAAN

### Summary

Two patients with serologically proven secondary syphilis also showed multifocal superficial thrombophlebitis. All manifestations cleared when appropriate antisyphilitic treatment was instituted. Although *Treponema pallidum* could not be demonstrated in the thrombophlebitic veins, the organism was considered responsible, either directly or indirectly. Multifocal superficial thrombophlebitis should be regarded as a new sign of secondary syphilis.

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Department of Dermatology, University of Stellenbosch and Tygerberg Hospital, Parowvallei, CP

H. F. JORDAAN, M.MED. (DERM.)

Syphilis is a disease with protean manifestations and truly lives up to its reputation of imitating many diseases. This is well documented in recognized textbooks.<sup>1</sup>

Two patients with secondary syphilis, which manifested with widespread superficial thrombophlebitis as an integral part of the clinical picture, are presented. In both cases the response to penicillin was dramatic.

The spirochaetemia of secondary syphilis leads to multi-system involvement, but multifocal superficial thrombophlebitis has not been described as a sign. A computer-based literature search on the MEDLINE database extending back to 1966 produced no references to this phenomenon. Possible pathogenic mechanisms are considered.

### Case reports

#### Case 1

A 23-year-old man presented at Tygerberg Hospital in 1981 with a 2-week history of painful bumps on the limbs and painless ones on the trunk. He was sexually active, but no history of a penile or mucosal lesion could be elicited. Before admission he had received salicylates and antihistamines without any effect. There were no other points of note in the history.

On examination a generalized microlymphadenopathy was evident. The glands were small and of rubbery consistency. There was a moth-eaten-type alopecia. A symmetrical maculopapular eruption of oval lesions 0,5-1 cm in diameter was present on the trunk. The eruption was more pronounced on the lateral aspect of the thorax.

On the limbs as well as on the upper anterior trunk there were numerous linear pigmented lesions varying from 1 to 12 cm in length. Their positions corresponded with the position of the superficial veins. On palpation these lesions were firm, but not overly tender and distinctly cord-like. Clinically they were typical of the healing stage of a thrombophlebitis. On the anterior thigh they manifested as ovoid pigmented nodules.

The VDRL test was positive to the titre of 1:256. The fluorescent treponemal antibody absorption (FTA-ABS) test was also positive.

A diagnosis of secondary syphilis was made and the patient treated with intramuscular benzathine penicillin G 2,4 million U weekly for 3 weeks. When he was seen at follow-up 2 weeks later, no lesions could be detected and after completion of treatment the patient was discharged.

A 2,5 cm segment of a subcutaneous thrombophlebitic vein was removed under local anaesthesia and submitted for histopathological examination. This confirmed the presence of thrombophlebitis without diagnostic features. A lymphocytic infiltrate occurred perivascularly, intramurally and in the thrombus. No spirochaetes could be demonstrated. An elastin stain demonstrated an internal elastic lamina but no external one.

This patient was diagnosed and treated for secondary syphilis on the grounds of bizarre alopecia, maculopapular syphilid, lymphadenopathy, thrombophlebitis and positive serology.

## Case 2

A 19-year-old man presented at the Department of Medicine at Tygerberg Hospital in 1981 with history of headache, malaise, anorexia and epigastric discomfort of 2 weeks' duration. He had previously been well. There were no other notable points in the history.

On examination he was pyrexial and had a generalized lymphadenopathy. The nodes were 0,5-1 cm in diameter, rubbery and non-tender. His throat was infected and the spleen was palpable.

A tentative clinical diagnosis of infectious mononucleosis was made. Numerous laboratory investigations provided no further clarification. The results of the following tests were negative: Paul-Bunnell (x 2); Weil-Felix; Brucella antibody test; PPD and blood cultures. The following were normal: chest radiograph; bone marrow; liver function and serum creatinine value. The white cell and differential counts were normal. The patient's erythrocyte sedimentation rate increased from 10 mm to 61 mm/1st h (Westergren) during his stay in hospital.

Eight days after admission the patient developed a skin rash and was referred to the Department of Dermatology. The rash consisted of 8-9 red nodular, cord-like, firm and tender symmetrical lesions on the anterior aspect of the thighs. Clinically they were consistent with a thrombophlebitis. On closer examination, several pigmented cord-like lesions were found on the lower legs. They were regarded as the healing stage of thrombophlebitis. A small scar was seen on the glans penis. The VDRL test was positive to a titre of 1:512. The FTA-ABS test was also positive.

A diagnosis of secondary syphilis was made and the patient was treated with intramuscular benzathine penicillin G 2,4 million U weekly for 3 weeks. His illness responded dramatically and at follow-up after 48 hours the lesions on the thighs had disappeared. Two weeks later no abnormality could be found on clinical examination.

A punch biopsy specimen of one of the nodular lesions was submitted for histopathological evaluation, but unfortunately the subcutaneous fat was not included in the specimen. The epidermis and dermis were normal.

This patient was diagnosed and treated for secondary syphilis on grounds of: constitutional symptoms, pharyngitis, generalized lymphadenopathy, healed chancre, thrombophlebitis and positive serology.

## Discussion

Two patients are described with secondary syphilis, both of whom displayed familiar clinical signs of the disease and the diagnosis was confirmed serologically. Additionally, both showed widespread superficial thrombophlebitis. This phenomenon has not been described previously. Institution of treatment with penicillin resulted in prompt healing.

Using the Warthin-Starry stain, *Treponema pallidum* could not be demonstrated in the thrombophlebitic vein from case 1. A fluorescent antibody technique was not used. Considering the fallibility of this stain, the possibility that few or no organisms were present, and the fact that histopathological examination was carried out during the healing stage, this finding is not unexpected.

The pathogenesis of thrombophlebitis is purely speculative. Direct lodging of spirochaetes in the wall of veins may give rise to inflammation and subsequent thrombosis. The spirochaetemia of secondary syphilis could provide the agent with access to vascular structures. A similar mechanism might be operative in rickettsial diseases. The affinity of *T. pallidum* for vascular structures is well known, but there is no explanation for preferential involvement of veins.

An indirect mechanism was considered as an alternative, possibly immunologically mediated.

The occurrence of thrombophlebitis in these 2 patients with secondary syphilis could be purely coincidental. The disappearance of all signs including the thrombophlebitis with appropriate treatment argues against this.

Thrombophlebitis is not an uncommon clinical diagnosis. However, the occurrence of widespread thrombophlebitis as a manifestation of secondary syphilis has not been documented previously. Clinicians are alerted to this interesting clinical presentation of this imitator of disease. Syphilis should be included in the differential diagnosis of multifocal thrombophlebitis.

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