Education

Éducation

Dr. Warren is Director of the History of Medicine Program, University of Manitoba, Winnipeg, Man.

CMAJ 1999;161(7):846-7

Osler's unusual case — Was it Churg–Strauss syndrome?

Peter Warren, MB

Ninety-nine years ago, on Oct. 15, 1900, William Osler presented a case to a meeting of the Johns Hopkins Hospital Medical Society. I will present the case in Osler's words[1] and then show that it has the features of what we now call Churg–Strauss syndrome. Osler's report began with the following paragraphs.

This is an unusual case in several respects. This young man came in on the 3d of October complaining of pain in the abdomen. His personal and family history are negative so far as this present condition is concerned. He had eaten abundantly of pork, and it is not known whether it was raw or cooked, as he is a Pole, and it is difficult to understand him.

His present illness began with a chill, accompanied by pain in the abdomen, and on the three following days he had nausea and vomiting. There was no diarrhea. There had been cough and expectoration since the onset of the illness and he had been confined to bed. On the night of admission the most remarkable feature noticed was a very deep cyanosis. The respiration was somewhat labored, being about 30 to the minute, but there was no urgent dyspnœa. There were numerous dry râles and much wheezing in the tubes. He remained in this condition of remarkable cyanosis with practically no fever, except a slight one on the third day; indeed, as a rule, his temperature has been sub-normal. The cyanosis was extreme and with it, which is noteworthy, he remained constantly recumbent.

On October 7, in addition to the cyanosis, petechiæ appeared over the body, first on the face and chest and then over the skin of the entire body except the legs. He presented a unique appearance, so far as our experience here is concerned, and looked very much like a case of malignant hemorrhagic smallpox. A differential count showed 11 per cent of eosinophiles. On the 8th of October he showed great tenderness of the muscles. The slightest touch on the muscles of the arms or legs caused him to wince. A portion of muscle was excised and showed marked degeneration with a great deal of fat in the fibres, but no trichinæ. On the 9th his leucocytosis rose to 52,000, the petechiæ had increased, his face was swollen, and he looked to be in a very critical condition. He was, however, rational, apparently comfortable and took his food fairly well. On the 11th the eosinophiles had risen to 25 per cent. Yesterday the cyanosis began to disappear. Cultures from the blood are negative and there is no Widal reaction. There is a trace of albumin and few granular casts.

No more clinical information is given. Osler went on to review the causes of cyanosis in general. In discussion, Dr. William Welch, newly elected president of the society, asked if there were any abnormal leukocytes, to which Dr. Futcher replied No, but that there were cells that were hard to classify as eosinophils or polynuclear leukocytes. Welch then reminded the society that the case was similar to black smallpox, in which it had been claimed that the leukocytic count is characteristic. No more was said, and the case was reported under the heading "Case of asthma with cyanosis, extensive purpura, painful muscles, and eosinophilia."[1]

What could this young man have had? At first sight, trichinosis seems likely, and certainly, as indicated by the opening paragraph of the case, Osler suspected it. Usually occurring after the ingestion of contaminated pork, trichinosis is caused by the nematode *Trichinella spiralis*. The larvae of the parasite migrate from the gut to the muscles and, to a lesser degree, to other tissues, including the lungs and the brain. Purpura and petechiae are rare. Osler was an acknowledged expert on trichinosis. He had been interested in the epizoa since his student days. In 1869, while still in Toronto, he had even studied their infiltration of the muscles as part of a routine dissection in anatomy. He published on trichinosis as early as 1876 and engaged in an exhaustive study of the parasites of the Montreal pork supply in 1883. In 1896 T.R. Brown, one of Osler's pupils, discovered the eosinophilia of trichinosis.[2] In 1899 Osler reviewed his personal experience with trichinosis, and the method for muscle biopsy, in the *American Journal of the Medical Sciences*.[3] Therefore, if Osler

had thought that the young man had an unusual presentation of trichinosis he might have been expected to say so, and the muscle biopsy should have confirmed it.

The differential diagnosis of trichinosis includes typhoid, eosinophilic leukemia and the vasculitides, in particular Churg–Strauss syndrome. Typhoid was excluded by the negative results of both culture and the Widal test. Leukemia was presumably excluded on the basis of Futcher's reply to Welch.

I suggest that Churg–Strauss syndrome would explain the clinical picture. This disorder, originally called allergic granulomatosis, allergic angiitis, was distinguished from periarteritis nodosa by Churg and Strauss,[4] who reported the presence of granulomatous lesions in vessels and connective tissues. The clinical picture is of a systemic vasculitis resembling polyarteritis (periarteritis) with 3 distinct features: a history of asthma, peripheral eosinophilia and pulmonary involvement in one-third to one-half of cases. The disease may involve various organs, including the gut and skin.[5] In their seminal paper Churg and Strauss reviewed 13 cases. They reported irregular fever in all cases and marked leukocytosis, as high as 60 000/mL, with the proportion of eosinophils reaching 84%. All patients had recurrent pneumonia, and heart failure was frequent. Abdominal pain and diarrhea were present in every case. As a rule, there was mild hematuria and albuminuria. In describing the skin lesions Churg and Strauss wrote "Of significance is the frequency of purpura (non-thrombocytopenic)." They did not observe myositis. Death was usually from cardiac or cerebrovascular disease. In the light of Osler's concern about trichinosis, it is of import that in 4 of the patients described by Churg and Strauss, the result of a skin test for *Trichinella* antigen was negative, but the level of precipitins was very high.

Further evidence that Osler was seeing a case of Churg–Strauss syndrome is given by a summary of findings in 16 cases by Lanham and colleagues[6] that was coupled with a review of 138 cases reported in the English literature up to 1982. These findings included asthma in all patients, pulmonary involvement in 70%, abdominal pain in 59%, purpura in 48%, cardiac failure in 47% and myalgia in 41%.

The râles observed by Osler may have been the result of pneumonia (a condition that can be interstitial) or cardiac failure. Both would have explained the cyanosis that perplexed Osler, but the patient's tolerance of the condition and his spontaneous recovery suggest that eosinophilic pneumonia was more likely. The wheezing may have resulted from asthma. Abdominal pain is common in the syndrome, but diarrhea less so. The renal involvement is usually benign and fits with the albuminuria and the low number of casts. Myalgia is common, and myositis occurs. Blind biopsy of muscle, as performed by Osler, was not recommended by Lanham and colleagues[7] because of its low diagnostic yield. Hence, Osler's nonspecific findings do not exclude the conclusion that the patient had Churg–Strauss syndrome.

I conclude that it is highly plausible that Osler's unusual case was one of Churg–Strauss syndrome. Osler's case report is an example of the scholarly contribution of a clinician who practises at the bedside and records carefully the observations made in this natural laboratory. Although Osler may have seen it first, he would have been the last to claim the syndrome to his name — he believed that those who fully describe a disease should get the credit. Churg–Strauss syndrome is a fitting eponym, for the studies of Churg and Strauss defined the disorder.

I gratefully acknowledge the help of Ms. L. Szczygiel, Osler Library of the History of Medicine, McGill University, Montreal, and the staff of the N.J. MacLean Health Sciences Library, University of Manitoba, Winnipeg.

References

1. Osler W. Case of asthma with cyanosis, extensive purpura, painful muscles, and eosinophilia. In: Proceedings of the Johns Hopkins Hospital Medical Society, October 15, 1900. *Johns Hopkins Hosp Bull* 1901;12:17.
2. Cushing H. *The life of Sir William Osler*. vol 1. Oxford: Clarendon Press; 1925.
3. Osler W. The clinical features of sporadic trichinosis. *Am J Med Sci* 1899;117:251-65.
4. Churg J, Strauss L. Allergic granulomatosis, allergic angiitis and periarteritis nodosa. *Am J Pathol* 1951;27:277-94.
5. Churg A. Pulmonary angiitis and granulomatosis revisited. *Hum Pathol* 1983;14:868-83.
6. Lanham JG, Elkon KB, Pusey CD, Hughes GR. Systemic vasculitis with asthma and eosinophilia: a clinical approach to the Churg–Strauss syndrome. *Medicine* 1984;63:65-81.

Reprint requests to: *Dr. Peter Warren, Director, History of Medicine Program, University of Manitoba, RS115 HSC, 810 Sherbrook St., Winnipeg MB R3A 1R8; fax 204 787-2420; warrencp@ms.umanitoba.ca*