medical history

Did Osler's Patient, Hannah W, Have Sarcoidosis?*
An Historical Footnote

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On March 30, 1894, an 11-year-old black girl, Hannah W, was admitted to the Johns Hopkins Hospital, Baltimore, complaining of swollen glands in the neck. William Osler was her physician. He examined the patient the next day and dictated the following note:

The child is well nourished and has good colour; the tongue is clean. The eyes are a little prominent. A remarkable feature is the symmetrical enlargement of the both parotid glands which stand out very prominently and tilt up the lobes of the ears. To the touch they are painless and have a firm, board-like hardness. The outlines and lobulations of the glands can be felt with the greatest distinctness. Both submaxillary glands can also be seen as a prominent nodular mass beneath the skin. The child's eyes are naturally prominent, but what adds strikingly to this feature is an enlargement of the lachrymal glands, causing marked bulging just above the outer canthus of each eye.

Further examination revealed generalized peripheral lymphadenopathy and splenic enlargement. The case fascinated William Osler. In the second edition of his textbook of medicine, published in 1895, he mentioned this case under the heading of "Chronic Parotitis." For the next few months, Hannah's condition remained stable. The salivary, lacrimal, and lymph glands did not change in size. On Oct 1, 1894, Osler noted: "The right parotid gland is now smaller than the left. The lachrymal glands have become somewhat smaller. The spleen is still to be felt nearly two fingerbreadths below the costal margin." The patient did not have systemic symptoms of fever, anorexia, or weight loss. Indeed, after studying the picture, which Osler published in the American Journal of the Medical Sciences, January 1898, one is struck by the bright-eyed, well nourished child who appears to be in no apparent distress (Fig 1). As we learn from the history, the patient was neither anemic nor did she have a significant leukocytosis. During January and February of 1895, the patient had a swelling of the nose and thickening of the nasal septum with distinct ulceration. The diagnosis of syphilis was entertained, and she was treated with potassium iodide and mercury.

The disease behaved rather strangely. Although the right lacrimal gland continued to get smaller, the left remained the same. She developed a sore throat and buccal ulceration. For the first time, on April 6, 1895, the patient became febrile and developed right-sided chest pain. Osler diagnosed it as an attack of pleurisy with effusion. She gradually improved and by July 19, 1895, her parotid, lacrimal, and submaxillary glands had disappeared. The spleen was barely palpable in September 1895. The patient remained well till April

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Figure 1. Eleven-year-old girl showing bilateral parotid enlargement. The patient appears to be in no acute or chronic distress.
18, 1897. She died in July 1897 of cavitary tuberculosis. On autopsy, there was no trace of the parotid glands, and the lacrimal glands were replaced by fibrosis.

Osler was intrigued by this patient because he was not able to pinpoint the cause of the clinical syndrome. Mikulicz had earlier described a 47-year-old man who had symmetrical enlargement of all the salivary glands and the lacrimal glands. He regarded the disease as a chronic infection. Kummel described a series of six cases, four men and two women, ranging from 23 to 47 years in age, with chronic enlargement of the salivary glands. In none of these cases was a definite diagnosis established. What, then, was wrong with Hannah W?

Did she have tuberculosis? Primary tuberculosis of the parotid gland is a rare entity.4 The lung, with its higher oxygen tension, seems to favor the growth of the tubercle bacillus, whereas the parotid gland does not. The infection usually starts in the tonsils or elsewhere in the oral cavity. About one third of the cases present in the second or third decade of life. Two clinical forms of tuberculous parotitis are recognized: an acute inflammatory infection with diffuse glandular involvement, and a chronic indurated mass with subsequent cold abscess and fistula formation. The diagnosis is made by appropriate staining and culture and the tissue examination. The history and physical examination here are not consistent with tuberculous parotitis. Osler thought of tuberculosis but dismissed it because the idea was “not confirmed by the subsequent history.” Furthermore, cultures made from the mucus squeezed from the parotid ducts were negative.

Did she have a fungal infection? Actinomycosis, perhaps more frequently than tuberculosis, affects the salivary gland. Usually the involvement is unilateral. The characteristic features are progressive inflammation with sinus tracts. This diagnosis can be safely excluded.

Allergic reactions are relatively common causes of parotid enlargement in childhood. Fish products and strawberry are frequently involved. The association of other allergic phenomena such as eczema, dermatitis, bronchospasm, and rhinitis support the diagnosis. The condition usually resolves rapidly and is recurrent. Although our case did have some swelling of the nose and thickening of the nasal septum, these features do not suggest an allergic cause.

Osler thought that the clinical picture was associated with inherited syphilis. The patient, however, had no evidence of secondary or tertiary syphilis. Unilateral or bilateral parotitis may occur in secondary syphilis, but is an unusual manifestation of the disease. We believe Hannah W. had systemic sarcoidosis.

In 1909, Heerfordt6 described a syndrome of parotid enlargement, uveitis and facial paralysis, but believed that the syndrome was due to mumps. Almost a quarter century later, Waldenstrom described five patients with bilateral parotid enlargement and uveitis. Four of them were women, all over 30 years of age. Epithelioid granulomas were observed in the parotid gland tissue of one patient. Thus, Waldenstrom is credited for recognizing parotid enlargement as but another manifestation of multisystem sarcoidosis. In a series of 388 patients studied by Greenberg et al.4 23 (6 percent) had enlarged parotid glands. The most frequent features were lymphadenopathy and splenomegaly. Lacrimal gland enlargement occurs in about 5 percent of sarcoidosis patients with parotitis.7 Sarcoïd parotitis may be asymptomatic or symptomatic (dry mouth), unilateral or bilateral, and acute (transient) or chronic (persistent). Osler’s patient most likely had multisystem sarcoidosis involving the parotids, lacrimals, peripheral lymph nodes, spleen, and the nasal mucosa. The disease ran a benign course and subsided within a two-year period. Fever and chest pains are reported in patients with sarcoidosis. The development of pulmonary tuberculosis, most likely, was not related to sarcoidosis.

Thus, William Osler perfectly described a case of multisystem sarcoidosis. The disease had subsided, and at autopsy no evidence was to be found of a noble disorder which eluded the noble physician. Nature had robbed Osler of an opportunity to describe a new disease.

REFERENCES

2. Osler W. Practice of medicine. New York: Appleton and Co. 1895