

9-1957

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Recommended Citation

Kavanagh, Doris (1957) "Quincke's Disease," *Henry Ford Hospital Medical Bulletin* : Vol. 5 : No. 3 , 176-178.
Available at: <https://scholarlycommons.henryford.com/hfhmedjournal/vol5/iss3/5>

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QUINCKE'S DISEASE

DORIS KAVANAGH M.D.*

Quincke's¹ disease has been known to the medical profession for at least 200 years. It was first differentiated from severe urticaria by Milton² in 1876 in a paper entitled "On Giant Urticaria". Six years later Quincke's paper on "Acute Circumscribed Edema of the Skin" attracted general attention. In it he presented his interpretation of the symptoms and the condition has since then borne his name. The term angioneurotic edema was introduced by Strübing³ in 1885 and has been the name most commonly used to designate the condition since that time. Osler⁴ in 1888 published an excellent description of the familial type — the least common form of angioneurotic edema. Until 1930 only 22 family histories had been described in the literature. However, since then many more such cases have been reported.^{5,6,7}

The condition first makes its appearance in childhood, (it has been reported in a 56 hour old infant⁸) and usually persists throughout life. It is characterized by recurrent transient swellings of the skin, mucous membranes, or viscera. When involving the skin there is usually a sharp, stinging or itching sensation followed within a few minutes to hours by swelling which may rapidly progress to alarming proportions. The area is pale, cool, non-pitting, and is accompanied by a feeling of fullness. Itching and pain are usually absent. There is a strong tendency to recurrence especially in previously affected areas. When it effects the mucous membranes of the gastro-intestinal tract, crampy abdominal pain, nausea and vomiting may occur often accompanied by transient anuria. Almost immediate death may occur from edema of the larynx. A case in point was described by T. W. Griffith, of a young woman so affected. He writes, "Suddenly she started up, apparently tried to speak, clutched at her own neck . . . got blue in the face and died almost at once."¹⁰ The viscera may also be involved. Crowder reported a case of an 18 year old girl with uterine involvement. Within 30 minutes the uterus "presented above the pelvis, as large, and to the touch very much the appearance of a 20 lb. cannon ball."⁹

Death commonly occurs from edema of the larynx. At autopsy the mucous membranes of the larynx are grossly edematous, pale, and translucent. The sides are usually in apposition, cutting off the airway. Some small amount of viscid mucous may be present. On microscopic examination the edema is found to affect the mucous membranes, deeper connective tissue, and even the substance of the muscles. The cells are chiefly of the mononuclear type. Occasionally the lungs have been reported affected, being voluminous and purple with pink frothy fluid exuding on pressure. The necropsy findings of such a case in a 14 month old child have been reported in detail by Wason.¹¹ Some patients with gastro-intestinal symptoms have come to surgery during the height of an attack. Two cases have been reported by Spaulding.⁵ One, a 53 year old male, had 50 cm. of grossly edematous jejunum resected. The other case presented a considerable amount of fluid in the abdominal cavity, as well as large edematous lymph nodes in the mesentery of the small bowel. Barnett⁶ reports of one patient operated for abdominal pain at McBurney's point, nausea and a leucocytosis

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Kavanagh

of 22,000; 90 per cent of the cells were polymorphonuclear leucocytes. At surgery the peritoneal cavity was filled with pink staining fluid which coagulated on standing. The appendix was normal.

The mortality rate in affected members has been reported between 16 per cent by Fairbank and 51 per cent by Crowder.⁹

Quincke's disease is transmitted as a dominant trait under the Mendelian Law. Those individuals who escape in an affected family are not likely to transmit the disease while those affected transmit it to approximately half of their children. The mean age of death, usually, from edema of the larynx has been reported as 34.4 years. Thus death early in the reproductive years may account in part for the infrequency with which the disease is seen. The affected families tend to die out. Factors precipitating individual attacks are usually unknown. Pressure or trauma to a part may precede an attack. Excessive alcoholic intake or emotional stress in many cases may precipitate swelling. One would like to demonstrate a sensitizing agent in these cases, but that has not often been found and skin tests with foreign proteins have not proved helpful. Rarely the removal of a focus of infection, e.g. tonsils, teeth, has seemingly afforded temporary relief, lending a little support to the bacterial sensitization theory.

In general the treatment of Quincke's disease has been unsatisfactory. A host of medications have been tried without changing the course of the disease. Some of the treatments used and found ineffective are adrenalin, ACTH, cortisone, thyroid extract, antihistamines, elimination diets, desensitization programs, histamine, sedatives, vitamins, ultraviolet rays, etc. Sheldon reports that intravenous benadryl tended to abort some attacks when given early, but in other instances in which it was tried it had no effect. Tracheotomy is always indicated in laryngeal obstruction, and in some patients it has been necessary to repeat this procedure four or five times for repeated attacks.

Case Presentation:

Mr. W. C. has been followed at the Henry Ford Hospital for many years. His first attack of swelling, involving the lips, occurred at the age 14. From that time on he had repeated attacks of swelling, usually involving the lips, hands or neck. At the age of 26 he had his first attack of laryngeal edema and since then this has recurred several times a year.

His last admission was occasioned by one of his typical attacks, and will be reviewed in detail. On January 7, 1957 we contacted him requesting that he come to the clinic for protein testing the following day. He agreed but began to worry about the procedure and drank twelve cups of coffee. By 10 P.M. he felt nervous and noted a small area of swelling in the sub-occipital region. He awoke at 4 A.M., January 8, 1957, in respiratory distress and came to the hospital. He denied any alcoholic intake for 4 months prior to the attack. (It had been our impression that previous attacks were related to excessive alcoholic consumption.) On admission he was noted to be a slightly obese, white male, very apprehensive and appeared his stated age of 43. His temperature was 98.6, pulse 90, and respiration 24. The tissues of the neck were markedly swollen and pale, and the overlying skin appeared tense. His voice was hoarse and he was in mild respiratory distress. Laryngoscopic examination showed a pale, boggy swelling of the soft palate, uvula and tonsillar pillars. The larynx on this occasion appeared normal.

Treatment consisted of adrenalin, intravenous aminophylline, and 100 mg of hydrocortisone given intravenously. The attack began to subside about 12 Noon; 8 hours after awaking in respiratory distress, and it was over in 12 hours.

The patient's family history for angioneurotic edema is very interesting. His paternal grandfather had attacks of severe gastro-intestinal cramping all his life. The grandfather had four

Quincke's Disease

children, 2 not affected, and 2 affected. The two affected members were the patient's father who had recurrent attacks of laryngeal edema and who died at the age of 55 of the disease, and the father's sister who also had attacks of laryngeal edema. The patient's father in turn had 2 children; the patient's brother, who is not affected, and the patient. The patient has 5 small children the oldest of which is 12. At present only one child seems affected, a 7 year old daughter with recurrent bouts of gastrointestinal distress, and swelling of the dorsum of the hands.

Summary:

(1) Available literature on the clinical entity known as Quincke's disease has been reviewed.

(2) A patient with Quincke's disease in which there is a strong hereditary pattern has been presented.

(3) Treatment for Quincke's disease to date has been quite unsatisfactory. We had hoped that this condition would be helped by the use of steroid preparations. To date reports in the literature on the use of steroids, and their use during 3 attacks in our patient have not been too encouraging. Further observation and treatment with these newer therapeutic agents would seem necessary before their true value can be ascertained.

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