OZENA AS A PART OF SYNDROMES

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Ozena may have become a less frequent disease than in earlier days, its treatment however, remains a problem. It is generally accepted that different factors may lead to the syndrome of the wide nose, which is mostly called ozena when foetor and Klebsiella ozenae are found and in absence of these two symptoms atrophic rhinitis.

Dependent on its etiology ozena can be divided into: 1. **primary ozena** when no external causative factor seems to play a role and 2. **secondary ozena** when the disease is a result of nasal or sinus surgery or due to chronic infection.

Primary ozena is most frequently found as a disease by itself and can then be called ozena simplex. Two types of ozena simplex can be distinguished: on the one side a type with internal nasal atrophy only, on the other side a type with both internal and external nasal atrophy.

Not always however ozena is a disease by itself. Sometimes it is found to be a part of a more general syndrome i.e. the ectodermal dysplasia syndrome and Sjøgren's syndrome.

Consequently the following classification of the different manifestations of ozena can be made.

I. Primary ozena

A. ozena simplex < internal atrophy
internal + external atrophy

B. ozena, part of. 1. ectodermal dysplasia syndrome
2. Sjøgren's syndrome

II. Secondary ozena

We will now consider these different types separately.

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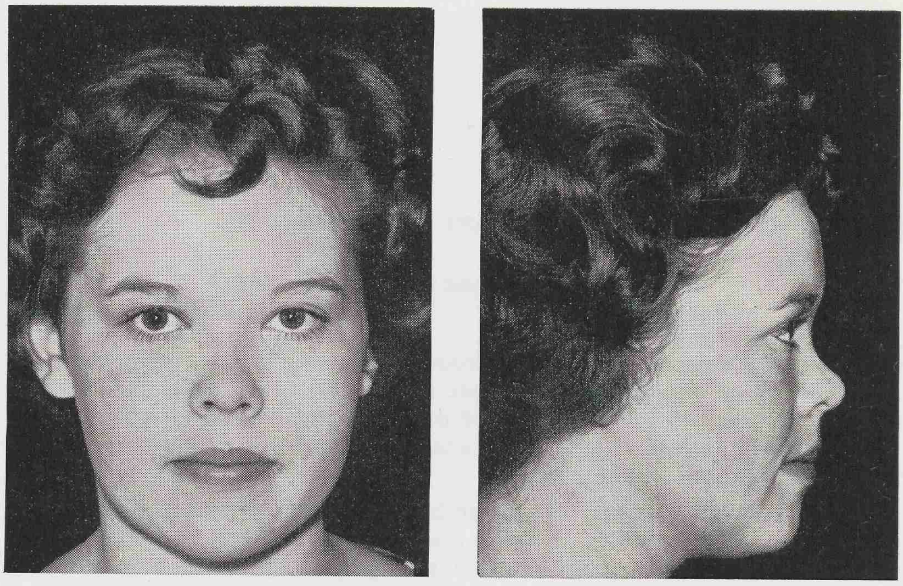


Fig. 1a and b: Shortness of the nose and small prominence of the dorsum in a girl with ozena simplex. Blood protein pattern disturbed.

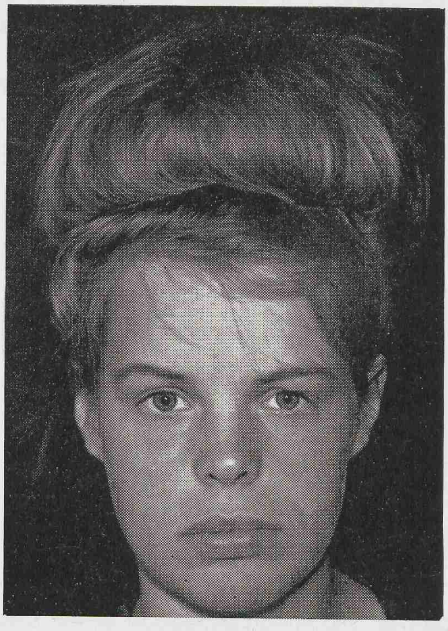


Fig. 2. Small short nose in a case of ozena simplex.

1. Ozena simplex with and without external nasal atrophy.

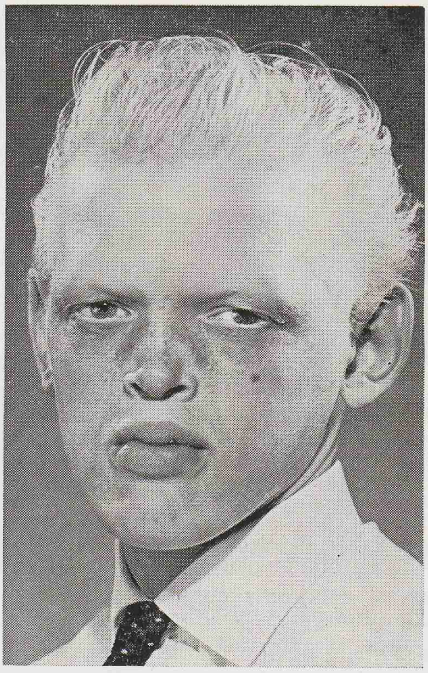
In the greatest number of patients with ozena an internal nasal atrophy is found only. Some of them however also present an underdevelopment or atrophy of the external nose and sometimes of the face or even of the whole body as well. Photographs 1a, b and 2 show patients with an ozena with both internal and external nasal atrophy. The height of the nose in both cases is too short in relation to the face and the prominence of the lobule and the cartilaginous and bony nasal pyramid is too small. There seems to be no relationship between the degree of the internal atrophy and the presence of an external nasal and facial atrophy. The patients who show an external atrophy however seem to have developed their ozena in early childhood. Therefore one is inclined to think more of an underdevelopment of the external nose and of the face rather than of an atrophy. The two varieties of ozena, the first type with internal atrophy only and the second type with both internal and external atrophy are not only met in cases of ozena simplex, but also in patients in which ozena is part of a syndrome, as will be discussed below.

2. Ozena as part of the ectodermal dysplasia syndrome.

Ectodermal dysplasia is a rare congenital syndrome consisting of hypotrichosis, anhidrosis, hypodontia and atrophic rhinitis/ozena. Other symptoms that may be present are nail dystrophies, malformation of the external ear, mental deficiency and diminished salivary and lacrimal secretions. This entity is best known by dermatologists because of the hypotrichosis and by pediatricians as to the inability of the patient to sweat which especially in children may induce a disregulation of body temperature and consequently high fever. The syndrome has also been described by rhinologists, e.g. by Christ (1913) and by Nager (1920). Its heredity was shown among others by Siemens (1937). Among the ozena patients who have been treated in our Department during the last few years, we have observed 4 cases with a complete or partial ectodermal dysplasia syndrome.

Patient A, a boy of 14 years of age presented the complete syndrome (fig. 3a-c). The hair of his scalp is scanty and non-pigmentated. The eyebrows and the hairs on the remaining part of his body are completely lacking. His skin is dry and pale and he is unable to sweat as was proven by different tests done in the dermatological Department (Verstege 1955). Lacrimal secretion is absent. The boy showed an ozena with external nasal and facial underdevelopment. Both the bony and cartilaginous vault are underdeveloped. The X-ray of his skull shows the internal nasal atrophy and the hypodontia (fig. 3c). Only 3 teeth in the lower jaw have developed.

Patient B, a man aged 32, showed the complete syndrome. He was seen in the Department of Dermatology and afterwards by us. There is the same type of hair, no development of teeth at all and an internal as well as an external atrophy of moderate degree (fi. 4a and b).



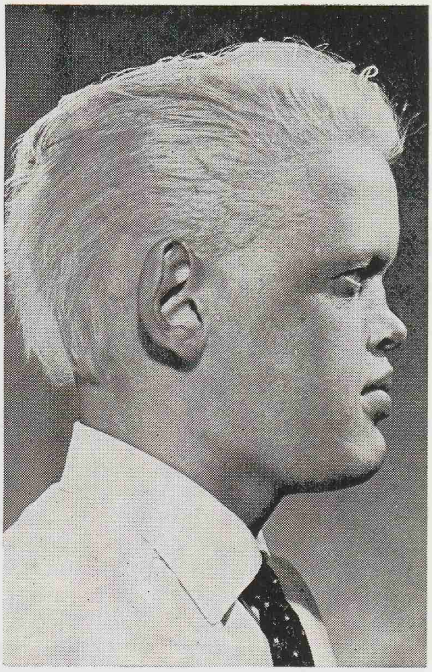


Fig. 3a en b: Patient A - Ectodermal dysplasia syndrome. Anhidrosis, hypotrichosis (note the absence of the eyebrows), hypodentia and ozena with external nasal and facial atrophy are present.

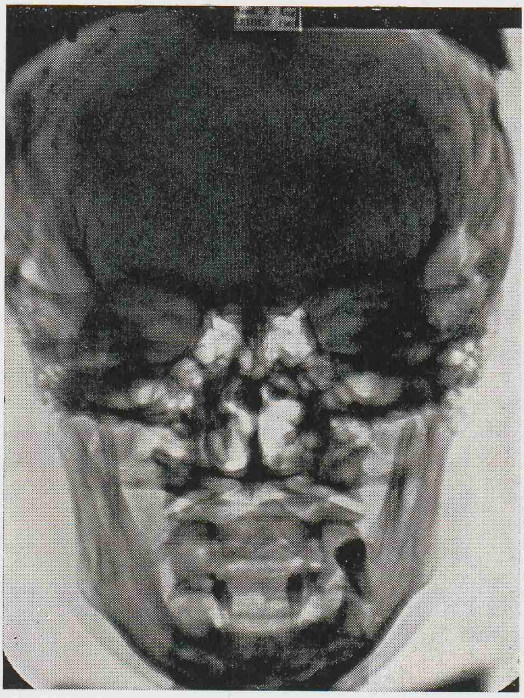
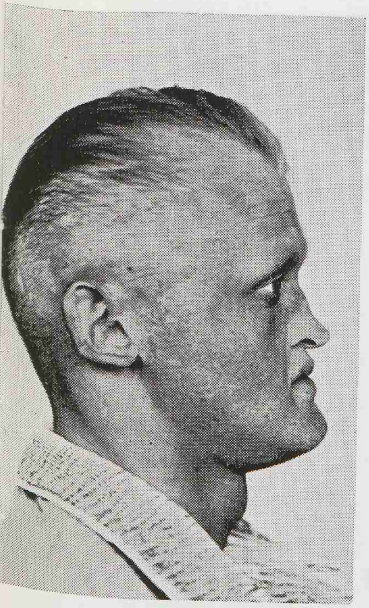


Fig. 3c. Spacious internal nose and hypodontia. Only two front teeth and one molar have developed.



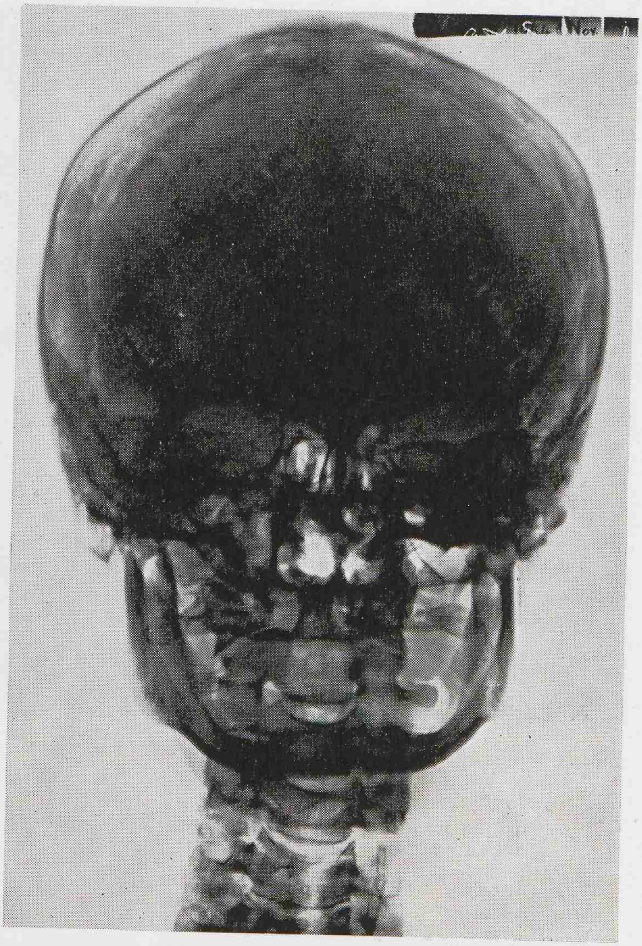


Fig. 4a en b.
Patient B - Anhidrosis, hypotrichosis (scarce thin hair on the scalp, no eyebrows), adontia and ozena with external nasal and facial atrophy.

Patient C, a man 28 years old showed the hypotrichosis, the hypodontia and an atrophic rhinitis. In his case too the hair of the scalp is very thin and without pigment (fig. 5). There was a marked internal nasal atrophy whereas the external nasal pyramid was normally developed. This patient only had two front teeth and now has a dental prothesis.

Patient D is a feeble minded girl of 22 years (fig. 6). In her case the combination existed of hypotrichosis, anhidrosis and atrophic rhinitis. The nasal underdevelopment or atrophy concerned both the internal and external nose and also the face to some extent.

In the ectodermal dysplasia syndrome all different combinations of hypotrichosis, anhidrosis, hypodontia, ozena with or without external nasal atrophy and some more rare malformations can be present. The possibility has to be considered whether a gradual transition from the complete ectodermal dysplasia syndrome via different incomplete syndromes to ozena simplex may exist.

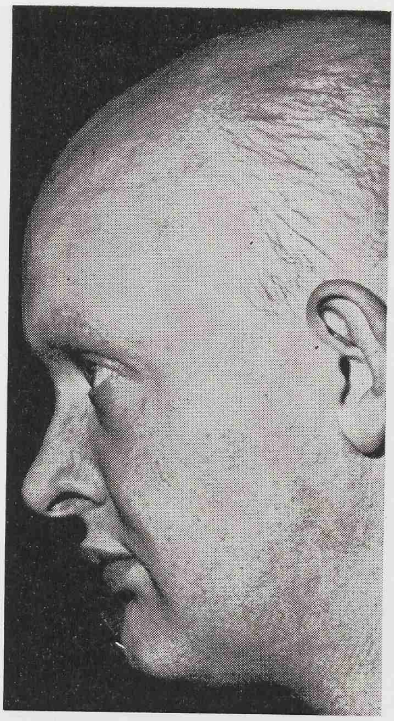


Fig. 5. Patient C - Incomplete ectodermal dysplasia syndrome: hypotrichosis (very few thin hairs on the scalp, no eyebrows), adontia and atrophic rhinitis. No growth disturbance of the external nose and face.

Fig. 6. Patient D - Girl with hypotrichosis, anhidrosis, mental deficiency and atrophic rhinitis with moderate sagging of the cartilaginous dorsum. Normal development of teeth.

3. Ozena as part of Sjøgren's syndrome.

Many of the symptoms of what now generally is called Sjøgren's syndrome have been described long ago. In 1933 they were put together by Sjøgren. In order to diagnose Sjøgren's syndrome the presence is required of:

— kerato-conjunctivitis sicca,

— xerostomia, sometimes accompanied by parotid swelling.

Often rheumatoid arthritis is also present and the sicca syndrome may be completed by atrophic rhinitis-ozena, atrophic pharyngolaryngitis, achlorhydria and atrophic vaginitis.

Just as in the ectodermal dysplasia syndrome often only some of these symptoms are present. All different combinations have been described. And here too, the question arises whether in relation to this syndrome ozena simplex is a single manifestation of a more general disease. A similar suggestion was put forward some years ago by Arslan c.s. (1960, 1963) and by Neuss

(1961). They wanted to include ozena in the group of collagen diseases. The argument of Arslan c.s. is the resemblance of the histopathology and of the disturbance of the blood protein pattern that would exist in ozena and in the collagen diseases. Some Italian investigators among others Salis (1956) and de Amicis and Corbetta (1960), have indeed reported a hypoalbuminemia and a hyperglobulinemia in ozena patients. At present however the value of the idea of grouping together different diseases as "collagen diseases" is questioned. Nevertheless it is still interesting to pay attention to the blood protein ratios as found recently by Bunim (1960) in different subgroups of Sjøgren's syndrome in relation to ozena.

In all his patients with Sjøgren's syndrome Bunim observed a lowering of the albumin fraction and an increase of the total globulin and of the gamma-globulin fraction. This was also found in patients, who showed the "sicca" symptoms (kerato-conjunctivitis, xerostomia, atrophic rhinitis etc.) only. These findings are therefore essentially the same as those reported by de Amicis and Corbetta in ozena patients.

In our Department blood protein determinations have been performed in a number of our recent ozena patients and the results have appeared to be normal in most cases. In three patients (women of 18, 18 and 59 years old) however a rather severe disturbance of the normal ratio was found i.e. a decrease of the albumin fraction and an increase of the globulin fraction. Serological tests (Latex fixation test and Rose-Waaler test) have always been found negative.

Summarizing we may say that primary ozena can present itself in several ways:

1. as internal nasal atrophy only;
2. as internal and external nasal atrophy;
3. as part of the ectodermal dysplasia syndrome;
4. as a symptom in Sjøgren's syndrome.

Different incomplete syndromes in between are possible. This suggests that primary ozena ought not to be considered too much as an isolated nasal disease and as a nosological entity. It possible is a manifestation of a more general disease. The presence of a disturbance in the blood protein pattern similar to that in Sjøgren's syndrome in some of the patients with ozena simplex is in favour with this assumption.

SUMMARY

Ozena may be a disease by itself (ozena simplex) or a part of a syndrome. Two types of ozena simplex can be distinguished: a. type with internal nasal atrophy only and a type with atrophy of both the internal and external nose and sometimes of the face too. Ozena/atrophic rhinitis can also present itself also as a part of the ectodermal dysplasia syndrome (anhidrosis, hypotrichosis, hypodontia and ozena) and of Sjøgren's syndrome (kerato-conjunctivitis sicca, xerostomia, rheumatoid arthritis, ozena e.o.) Four personal cases of ectoder-

mal dysplasia syndrome are discussed. In both the ectodermal and Sjøgren's syndrome all different combinations of the symptoms appear to be possible. The question is raised whether a gradual transition from ozena simplex to the two complete syndromes may exist. Consequently ozena has not to be considered too much as an isolated nasal disease but as a more general disease. This suggestion was made before by other authors because of certain histologic observations and abnormalities of the blood protein pattern found in ozena patients. The existence of a disturbed blood protein was confirmed in a few of our ozena simplex patients. The abnormalities were similar to those in Sjøgren's syndrome. Serological tests however were negative.

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