

A NEW FINDING IN ORAL CAVITY IN PSEUDOXANTHOMA ELASTICUM

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ABSTRACT

We made precise observations of the presence of symmetrical lesions especially in the mucous membrane of the oral cavity, based on the view that pseudoxanthoma elasticum is in general a systemic disease of elastic fibers and that similar lesions to those present in the skin are very likely to occur symmetrically in other regions. As a result, we found yellowish-white enanthemata of the same appearance at symmetrical sites of the hard palate in both of our latest 2 cases. The degeneration of elastic fibers, quite similar to the findings of the skin, revealed by histological observation of the lesions, together with remarkable calcification demonstrated by Kossa's stain, led to the conclusion that the enanthemata are those of pseudoxanthoma elasticum.

We report our findings here as we believed that the enanthemata which occurred in identical regions and with the same appearance and histological findings, even though in only 2 cases, might probably be due to pathological change in the mucous membrane of the oral cavity which is characteristic of this disease.

Pseudoxanthoma elasticum, a disease resembling xanthoma clinically, was first described in 1884 as another type of xanthoma by Balzer who examined patients with xanthoma-like papule associated histologically with regressive degeneration of elastic fibers. Later, in 1896, Darier¹⁾ who made a detailed histological investigation of similar diseases, differentiated the disease from true xanthoma based on the absence of xanthomatous cells in the former, and named it pseudoxanthoma elasticum.

Other synonyms for the disease include xanthoma elasticum (Balzer), navus elasticus (Gutmann), Elastom der Haut (Juliusberg) and pseudoxanthoma elastoclasium calcosum (Ohno). The disease is not quite rare since more than 100 cases have been reported both abroad and at home. However, a papule of the mucous membrane has very rarely been found.

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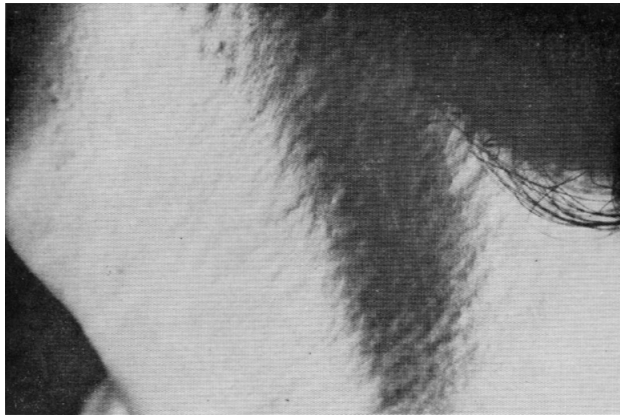


FIG. 1. Patient with pseudoxanthoma elasticum, showing lesions on the left side of the neck

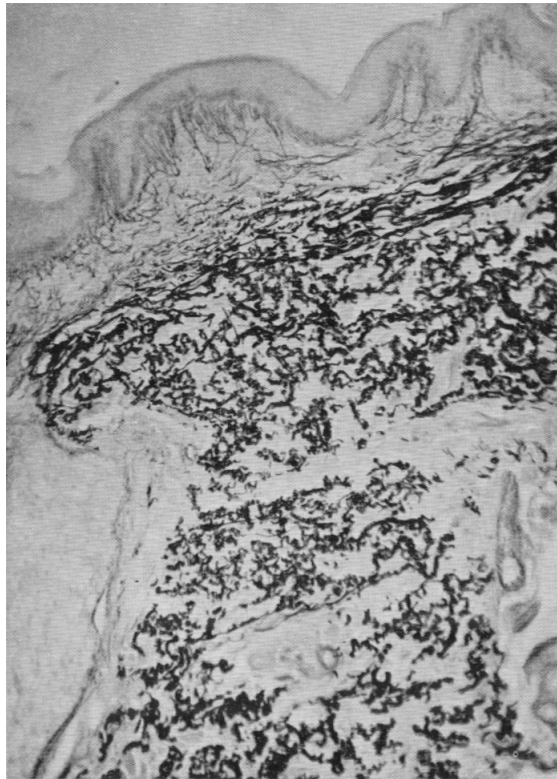


FIG. 2. (Weigert's stain)

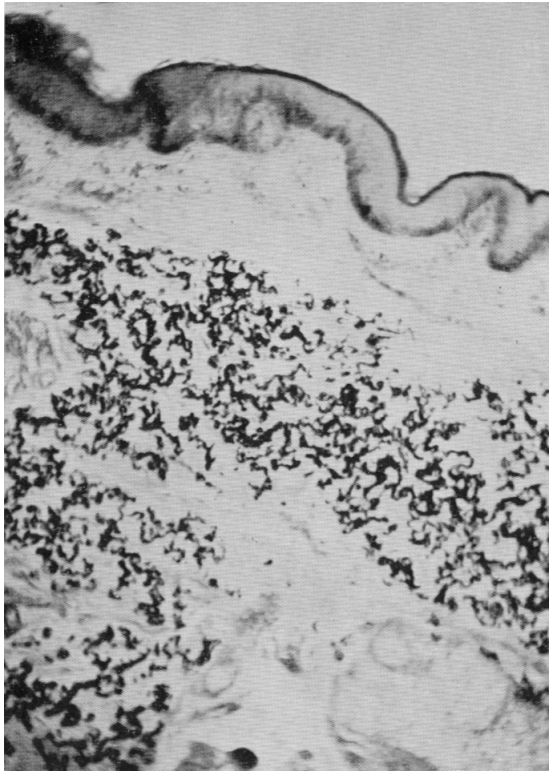


FIG. 3. (Kossa's stain)

Herein is described a highly characteristic finding obtained recently by us in the oral cavity of 2 cases of pseudoxanthoma elasticum, the finding which may be obtained in similar other cases by anyone who will take the patients to examine with care.

Case 1: A 20-year-old female

Family history: The parents of the patient are half-blood brother and sister, and 3 of her 7 brothers and sisters are deaf and dumb.

Previous history: none to be mentioned.

Present history: Yellow exanthemata seen on both sides of her neck since she was about 15 years old. She visited us as the exanthemata which had been left untreated due to absence of subjective symptoms recently began to extend.

Findings of the skin: numerous round or elliptical yellow papules, the size of a stick needle's head, were seen like seeds sown on both sides of the neck, in the armpits and in the groin. These were found to fuse into a re-

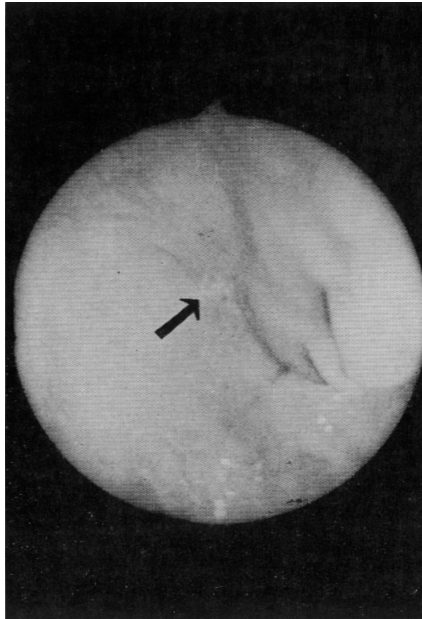


FIG. 4. Enanthemata on hard palatal region (left side)

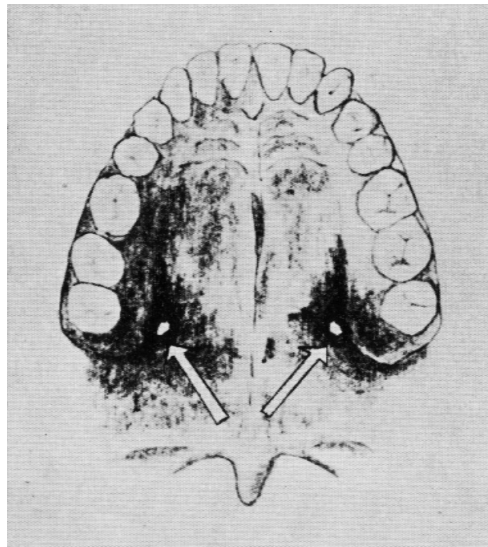


FIG. 5. Shema showing situation of enanthematas

ticular form (Fig. 1).

The histological findings of the skin eruption consisted of degeneration of



FIG. 6. (weigert's statn)

elastic fibers in the middle and deep layers of the corium revealed by staining by Weigert's method: it was found that the elastic fibers either swelled irregularly and severed short, or were strangulated to show disorderly arrangement, or formed knots or lumps. Kossa's stain also revealed calcification, with degenerated elastic fibers dyed black. This calcification is characteristic and is one of the important findings in the histological picture of the disease (Fig. 2 and 3).

Diagnosis: Pseudoxanthoma elasticum

Findings in the oral cavity: Yellowish-white enanthemata, the size of red beans, were found wymmterically on both sides of the hard palatal region, on the inner aspect of molar III and inner and posterior sides of the alveolar process (Fig. 4 and 5).

The histological findings of the enanthema consisted of the degeneration of elastic fibers in the deep layer of the hard palatal mucous membrane revealed by Weigert's stain: the elastic fibers were found to swell irregularly, severed short and arranged irregularly. Kossa's stain also revealed calcifica-

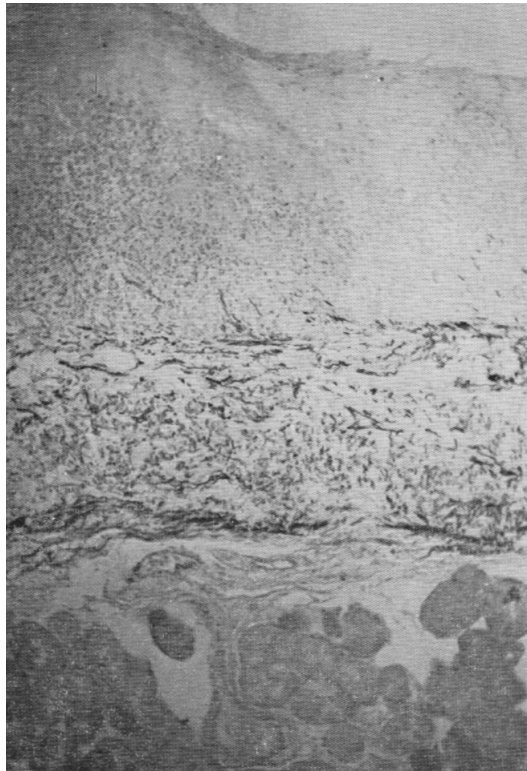


FIG. 7. (Kossa's stain).

tion, with degenerated elastic fibers dyed black. It was established from these findings that these enanthemata seen symmetrically on both sides of the hard palate represent pseudoxanthoma elasticum the same as the papules on the skin (Fig. 6 and 7).

Case 2: A 19-year-old female

Family history: None to be mentioned.

Previous history: Not marked as patient is constitutionally healthy.

Present history: She found yellow exanthemata on both sides of her neck and in the armpits since she was about 12 years old, which were left untreated as no subjective symptoms developed. She visited us when the lesions began to extend.

Findings of the skin: On both sides of the neck and in the armpits, chelidon and groin, numerous red bean-sized round or elliptical yellow papules were found disseminated, some of which were found to fuse into a reticular form.

The histological findings of the skin eruption were almost identical with those of case 1.

Diagnosis: Pseudoxanthoma elasticum

Findings of oral cavity: As in case 1, yellowish-white enanthemata, the size of red beans, were found symmetrically on both sides of the hard palatal region, on the inner aspect of molar III and inner and posterior sides of the alveolar process. The histological findings were also the same as in Case 1, and the patient was diagnosed as pseudoxanthoma elasticum.

DISCUSSION

Our 2 cases of pseudoxanthoma elasticum belong to what has been called *typus Darier-Balzer* by Friedmann²⁾ (1921), which occurs mainly in younger people and more frequently on both sides of the neck and other regions symmetrically, with the lesion protruding a little above the surface of the skin and of a xanthoma-like yellow color and, histologically, presenting circumscribed degeneration of elastic fibers mainly in the middle or deep layer of the corium.

The age of onset is frequently unknown because subjective symptoms are hardly present. About 80% of patients, however, have been reported to be less than 20 years of age in our country and in our 2 patients also the disease developed at the ages of about 12 and 15 respectively. The disease occurs in greater frequency in women than in men, and our 2 cases were both females.

Throne-Goodmann³⁾, Hartung⁴⁾, Matras⁵⁾ and others reported that the disease was seen in people born of consanguineous marriages, and one of our 2 cases was also such a patient, so that it cannot be denied that a hereditary factor is closely related to the occurrence of the disease.

It is noteworthy that the papules on the skin frequently occur symmetrically, and the incidence of the disease at various sites in decreasing order is: the neck, armpits, abdomen, groin, chelidon and thorax. In our cases also the main lesions were found symmetrically on both sides of the skin of the neck, the most favorite site of occurrence.

The majority of patients complain of no subjective symptoms except occasional itchings. It is said that the skin eruption is generally a round, elliptical or rather polygonal nodule, the size of a pinhead or tonsil, which is present in numbers bulging slightly from the surface of the skin, while the papules are isolated and scattered or gather or fuse together into a linear, cordal, reticular or spotted pattern, some of which presenting lesions larger than the palm.

The exanthema is yellow like xanthema, distinctly demarcated and fairly hard, and usually reduces the elasticity of the skin. Incidentally, the surface of individual papula is smooth with no desquamation.

The histological findings of the disease are characterized by little changes in the epidermis and papilla, a significant change in the middle and deep layers of the corium and changes in the elastic fibers of the lesion—viz degeneration and deformation. Of essential importance is change that has been termed elastoclasia and elastorrhexis by Darier¹⁾.

As to the etiology of the disease, Groenbald⁶⁾ stated that it may occur as the result of various functional disorders around puberty in those who have inherited a disposition to nutritional disturbance of elastic fibrous substance not only of the skin but of the body in general. Our finding agrees with his opinion. In a word, the disease is a type of systemic disease which may be attributed to primary regressive change of elastic fibers.

It has very rarely been reported that these lesions are also seen in the mucous membrane, a region other than the skin: They were observed in the mucous membrane of the oral cavity or throat by Darier¹⁾, Ramel, Lewis-Clayton⁷⁾, Matras⁵⁾, Foester⁸⁾, Urbach-Nekam and Kerl⁹⁾, and of the nose by Lewis-Clayton. None of these reports, however, suggested that the enanthema might be peculiar to the disease.

We made precise observations of the presence of symmetrical lesions especially in the mucous membrane of the oral cavity, based on the view that the disease is a systemic one of elastic fibers in the body in general, and believing that lesions similar to those present on the skin are very likely to occur symmetrically in other regions. As a result, we found yellowish-white enanthemata of like appearance at the symmetrical sites of the hard palate in both of our newest 2 cases. The degeneration of elastic fibers, quite similar to the findings of the skin, was revealed by histological observation of the lesions, together with a remarkable calcification demonstrated by Kossa's stain, and we were led to the conclusion that the enanthemata are those in pseudoxanthoma elasticum.

We reported our finding here as we believed that the enanthemata which occurred in exactly the same regions and with the same appearance and histological findings, even though in only 2 cases, might probably indicate a pathological change in the mucous membrane of the oral cavity which is characteristic of the disease. We believe that the same findings will be obtained in other cases of pseudoxanthoma elasticum of the skin by anyone who will take the pains to examine them with more care.

CONCLUSIONS

We found yellowish-white enanthemata symmetrically on both sides of the mucous membrane of the hard palate in 2 of our latest cases of pseudoxanthoma elasticum.

We believe that the enanthemata are probably characteristic of the dis-

ease and that the same finding is very likely to be obtained in many other cases if they are observed with care.

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