SYRINGOMA — A CASE REPORT WITH REVIEW OF LITERATURE

*S. P. AMIN AND †B. H. SHAH

Summary

A middle aged female patient having lesions of syringoma on face and scalp is reported with review of literature.

The available literature on syringoma in India is very scant and the incidence of it is so low that it is hardly ever thought of in differential diagnosis. The authors came across such a case which was missed clinically and diagnosed pathologically.

Case Report

A thirty year old married woman belonging to a low socioeconomic class was first seen in our outpatient department in March 1971. Her chief complaint was eruptions on the face of 4 years duration.

Physical examination revealed multiple, small, waxy, white, soft papules on her face (Fig. 1). The lesions were in hundreds and were round or oval in shape and varied from 1 to 3 mm. in size. They were discrete with little tendency to grouping and were limited to face and scalp. There were no associated skin lesions elsewhere on the body.

Fig. 1

Characteristic lesions of syringoma on the face.

The lesions had appeared before four years. Initially she had noticed two papules on the forehead. In the next two years, her whole face was involved; first the right side and then the left side. The lesions since then persisted without any change in size, colour or number. There was no effect of menstruation on lesions and seasonal variation was absent. The lesions were asymptomatic. The patient considered her lesions cosmetically unacceptable but still she was not eager to get them

* Registrar' Skin and V. D. Department;

† Honorary Skin and V. D. Specialist; Civil Hospital, Ahmedabad.

Received for Publication on 27—10—1972

removed because of her social status. Twice she attempted to get rid of the lesions by household measures with no avail. Once she applied common salt and at the other time she rubbed her face with coin. Her brother aged forty years is having similar lesions on the face for the past ten years.

The biopsy of the lesions revealed the typical histologic findings of syringoma (Fig. 2).

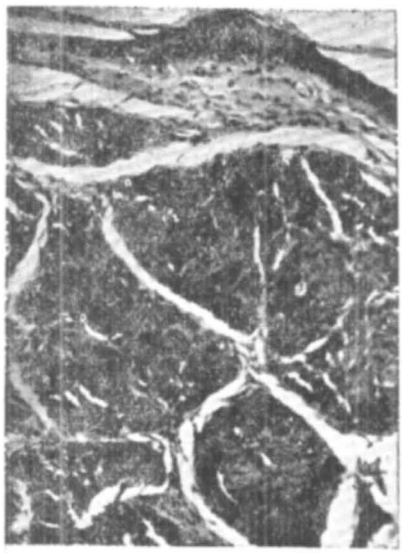

Fig. 2

Showing histologic changes of syringoma.

The patient was seen again in September 1972. The lesions were found to be stationary. The lesions then were treated by electrocautery.

Review of Literature

The apocrine gland in man is a vestigeal organ. In lower animals like apes and dogs they are present over the entire skin surface, serving a protective function. They are scent elaborating glands, giving a highly odoriferous and distinctive smell which aid in protecting the animal from attack.

With the loss of hair over the large areas of the body, there has been concomitant loss of apocrine sweat glands in man. A good example of ontogeny recapitulating phylogeny is seen in human embryo, where the abortive attempts at the production of apocrine sweat glands over the entire skin surface are found early in the fetal life. These initial apocrine buds regress and in full term foetus apocrine structures are seen only in limited regions. In man they are found over the axillae, the areolae of the nipples, the periumbilical region and the perianal and the genital regions. They may be also found anywhere on the body in atrophic forms. From evolutionary standpoint the apocrine glands stand midway between the sebaceous glands and the eccrine sweat glands. Its method of secretion is primitive but highly developed than that of the sebaceous glands.

There are three types of apocrine epitheliomas--syringoma, cylinderoma and myoepithelioma.

In syringoma differenation is directed mainly towards apocrine duct cells and in myoepithelioma mainly towards apocrine myoepithelial cells.[1]

These tumors are important mainly because they are frequently confused histologically with neoplasms of more serious prognosis, particularly with the neoplasms metastatic to cutis.[2]

Synonyms

Syringocystadenoma--Toerack.

Hyderoadenoma Eruptifs--Darrier & Jacquet.

Lymphangioma tuberosum multiplex-Kaposi.

Hemangioendothelioma Tuberosum multiplex--Jarish.

Nevus Epitheliomatosis Syringomatosus.

Nevus Syringoadenomatosis papilloferus.

Spiroadenoma.

Syringocystadenoma.[3]

History

In 1872 Kaposi and Biesiadeki reported the first case of Syringoma as 'Lymphangioma Tuberosum Multiplex'.

In 1887 Jacquet & Darrier presented a variation under the title of 'Hydradenomas Eruptives'.[4]

Darrier recognised two types of syringomas and called them Hideradenoma of lower eyelids and Eruptive hideradenoma.[5] Since then many cases involving diverse areas have been described.

Aetiology

In the past there were various schools of thought regarding its origin.

1. Origin from lymphatics or its excretory ducts.
2. Epithelial genesis.
3. From fully developed sweat glands or its excretory ducts.
4. Disturbance in embryonic coil gland cells.

At present by histochemical and electronmicroscopic studies it has been confirmed that the tumor does not arise from mature skin appendages but from immature pluripotent cells.[3]

Incidence

It is more common in women in their adolescence, especially in laundry workers. The lesion tend to progress in hot weather and regress in cooler weather.

Very early onset since birth or infancy as well as very late onset, past forty years have been recorded. Familial occurrence such as in two sisters, father and daughter, brother and sister, grandfather and grand-daughter are quoted.

The eruption appears in crops during first 2 to 3 years. They either remain indefinitely with minor fluctuations or disappear entirely.[2,6]

PATHOLOGY

Macroscopic

They are small, soft, slightly yellowish nodules, round or oval in shape and pin head size. They are frequently angular and flat topped. Often the typical tadpole shape may be seen. They may be present in hundreds. Individual lesions are often aligned in straight line along the cleavage line of the skin.

There may be associated skin lesions like nevi, fibrous angiomas, etc.[3,7]

Microscopic

Numerous small cystic ducts are seen in the dermis. Walls of the ducts are lined by two rows of epithelial cells which are flat. Occasionally inner row of cells show active secretion. Lumina is filled with colloidal material. Some cystic ducts possess small comma like tails of epithelial cells-tadpole appearance. They may also contain solid strands of epithelial cells in the corium. Occasionally rudimentary hair structures are seen. Trichoepithelium may be associated. Cellular heaps and strands are interspread between the cysts which are filled with clear material. Sometimes epithelial cells are keratinised and form horny cysts. Sometimes granules similar to keratohyaline bodies may be seen. Giant cells may be observed in the vicinity of the horny cysts. Epidermis may be

normal or thinner and displacement of the retepegs may be seen. In corium signs of displacement, rarefaction of the elastic fibres and increase in the number of connective tissue cells may be seen.

Evidences to confirm apocrine genesis.

1. Presence of active secretion.
2. Its simultaneous occurrence with rudimentary hair structures or trichoepithelioma.
3. Its appearance at puberty when apocrine glands first begin to function.
4. Prevalence of the lesions in areas of the skin where apocrine glands occur or used to occur phylogenetically.[3]

Electronymicroscopy

The tumor consists entirely of ductal structures, largely intraepidermal in type. Ductal cells boarding the lumina show microvilli and a band of periluminal tonofilaments. Numerous lysozymes are present in the cytoplasm of the periluminal cells and some periductal cells. Intracytoplasmic lumen formation characteristic of the immature intraepidermal eccrine duct is also observed. Keratohyaline granules in luminal cells are often observed. Since these two features are regularly observed in the upper portion of the intraepidermal eccrine duct, it can be concluded that differentiation of syringoma is towards this structure.[5]

Histochemistry

Histochemical investigations have been useful in determining the histogenesis because immature cells of cutaneous appendages of the human embryo contain certain enzymes that are quite specific for each cutaneous appendage.

Histochemical examination of the syringoma show a strong reaction for amylophosphorylase. Moderate to strong eccrine type of enzymatic reaction were obtained in syringoma such as malic or succinic dehydrogenese and leucine aminopeptidase. Acid phosphatase and B-glucuronidase the so called apocrine type of enzymes were occasionally demonstrated in periluminal areas. These areas on electron microscopy showed numerous lysosomes which are known to contain these enzymes. Thus by histochemical findings it may be concluded that in syringoma the differentiation is in the direction of eccrine structures.[5]

Clinical Features

Small skin coloured to yellowish papules are seen, frequently angular and flat topped with typical tadpole shape.

They are infrequently present over eyelids, neck, chest, abdomen, pubic regions.[8]

Cases involving thighs, knees, legs, even vulva[4] and penis[9] have been reported.

When situated on the trunk they usually diminish in number caudally.[3]

Eruptive Hideradenomas of Jacquet and Darrier with lesions arising in larger number in successive crops on the neck, chest, axilla, abdomen, mons pubis, and groin are rather rare.

Subjective symptoms are rare and often absent. Pruritus in warmer months and during menstruation is recorded.[6]

Differential Diagnosis

1. Fox Fordyce disease–Lesions are smaller more conical, periodically very pruritic.
2. Small epithelial cysts when multiple–They are fewer more yellow, often show one or more lesions with a characteristic central punctum.

3. Senile Angiomas–They are usually brighter red in colour.
4. Condyloma Accuminatum–They are multiple bilateral lesions with distinctive clinical features.
5. Epithelioma adenoides cysticum–The lesions are discrete, small, rounded, smooth, shiny, firm, papules. They are usually flesh coloured.
6. Milia–They are whiter, firmer, and can be easily enucleated.
7. Xanthomas–They have sulphur yellow colour.
8. Papular syphilides–The papules are coppery red, round, infiltrates 3–10 mm. in diameter. Serological tests for syphilis is positive.
9. Multiple sarcoids–Here the nodules are without characteristic distribution, firm, pale, yellowish, present for months or years.
10. Hydrocystoma–They are rounded or oval, pinhead to lentil sized lesions, tightly filled but elastic, translucent bladder like.[10]
11. Lichen Planus-[9] They are pruritic, small, flat-topped, polygonal red or violet coloured papules, centrally umbalicated. The lesions commonly occur on wrist, legs, glands penis & mucous membrane of the mouth.[9]

Complications

It is considered to be a benign tumor with no serious complications. Freeman and Winkelman had recently two cases of Basal cell carcinoma with eccrine differentiation in which features of syringoma were observed.[10]

Treatment

1. Light electrodessication of small individual lesions may be tried[10].
2. Superficial dermoabrasion – small lesions may be treated with it and patient may improve with one or two dermoabrasion[11].
3. Surgical excision for cosmetic reasons may be tried.[10,11]
4. The lesions may regress with atropine[2].

Acknowledgment

We are thankful to Dr. B. D. Mishra, Superintendent, Civil Hospital for allowing us to use Hospital records. We are also grateful to the patient for her co-operation.

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