Elastofibroma dorsi

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SYNOPSIS Two cases of elastofibroma dorsi are reported and the light and electron microscopic features analysed. The appearances suggest that the characteristic fibres and globules that stain as for elastin arise by denaturation of collagen. The fibres differ from the elastin of arterial wall, elastotic degeneration of skin, and pseudoxanthoma elasticum. The morphology and clinical behaviour indicate that elastofibroma dorsi is a reactive fibromatosis rather than a true neoplasm.

Elastofibroma dorsi is a tumour-like lesion, first described by Jarvi and Saxén (1961) who briefly reported four cases. Since that time a further seven cases have been reported, two additional cases have been seen in this hospital and are reported here (Table I), and Mackenzie, Wilson, and Cooke (1968) have seen a further three cases. With one exception, all the reported tumours have been characteristic in their site and morphology. The exception was a

the reaction was abolished by digestion in elastase for 75 minutes (Stemmermann and Stout, 1962). The two cases seen in this hospital have enabled the ultrastructure of the lesions to be examined. Comparison is made with elastic fibres of arterial wall and 'elastotic degeneration' of the skin.

CASE REPORTS

CASE 1 M.J., a white female clerk, age 61, presented with a mass below the right scapula which she had noticed for two months. It was hard but painless, and had increased in size since it was first noticed. On examination, the only abnormality was a firm, smooth mass 10 × 7 cm not attached to skin, but fixed to the serratus anterior muscle. Radiology showed a soft-tissue swelling without bone involvement. At operation a fibrous tumour was found under the lowest fibres of the trapezius muscle, deep and lateral to the lower border of the scapula to which it was fixed. On its deep aspect the tumour was attached to the periosteum of the ribs. Excision of the tumour and the lower border of the scapula was uneventful and there was no recurrence in the following 12 months.

Macroscopically the tumour consisted of an irregular mass of fibro-fatty tissue measuring 9 × 7 × 2 cm densely adherent to striated muscle. There was no capsule and the outline of the tumour was ill defined. Two smaller fragments of tissue were taken from the lower border of the scapula with adherent muscle, but they showed no evidence of tumour.

CASE 2 I.R., a white housewife, age 55, first noticed a lump the size of a half grapefruit at the inferior angle of the right scapula, six weeks before attending hospital. There was a four-year history of goitre confirmed as Hashimoto's thyroiditis and myxoedema controlled by thyroid extract. Examination confirmed the presence of a smooth firm tumour adherent to the scapula. At operation the tumour was found adherent to the periosteum of the sixth and seventh ribs and to the inferior angle of the scapula. Excision was incomplete but there was no evidence of recurrence three-and-a-half years later.

TABLE I
REPORTED CASES OF ELASTOFIBROMA

Reference	No. of Cases	Age (yr)	Site
Jarvi and Saxén (1961)	4	'Aged'	Lower subscapular space, one bilateral
Stemmermann and Stout (1962)	2	66	Subscapular
Delvaux and Lester (1965)	1	70	Bilateral subscapular
Barr (1966)	3	62	Subscapular
		45	Bilateral subscapular
		62	Subscapular
		75	Hip
Brown, Clearkin, Nakachi, and Burdick (1966)	1	58	Subscapular
Tighe <i>et al.</i> (1967)	2	61	Subscapular
		55	Subscapular

tumour described by Barr (1966) which occurred in the wall of a bursa overlying the greater trochanter of the femur. All the remaining tumours occurred in the interscapular or subscapular region, deep to the latissimus dorsi and rhomboid muscles. The appearances were those of fibro-fatty tumours often adherent to the scapula and to the underlying chest wall. The characteristic histological appearance was of many irregular deeply eosinophilic fibres and globules, 1 to 30 μ diameter, scattered in mature fibrous tissue. These eosinophilic fibres reacted as for elastin with Verhoeff's and Weigert's elastic stains and

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FIG. 1.

FIG. 1. *Eosinophilic fibres and globules characteristic of elastofibroma. Haematoxylin and eosin. $\times 540$.*

FIG. 2.

FIG. 2. *Elastofibroma. Elastic fibres have a central core and irregular borders giving a serrated outline. Weigert's elastic stain. $\times 540$.*

FIG. 3.

FIG. 3. *Elastofibroma. Numerous elastic fibres and globules of the elastofibroma. Verhoeff elastic-van Gieson. $\times 130$.*

FIG. 4.

FIG. 4. *Elastofibroma. A similar area to Fig. 3 after digestion in elastase for 75 minutes. Elastase-Verhoeff-van Gieson. $\times 130$.*

FIG. 5.

FIG. 5. Electron micrograph. Elastofibroma. Irregular elastic globules surrounded by typical collagen fibres. $\times 9,300$.

FIG. 6.

FIG. 6. Electron micrograph. Elastofibroma. Granular elastic globule extending between collagen fibres. $\times 30,000$.

Macroscopically the specimen consisted of fibro-fatty tissue in which was embedded a firm white fibrous tumour measuring $15 \times 10 \times 2.5$ cm without a recognizable capsule.

LIGHT MICROSCOPY

The histological appearances of both tumours were characteristic. They consisted of mature fibro-fatty tissue without a recognizable capsule. Confined to the fibrous tissue were many eosinophilic fibres and globules (Fig. 1). The fibres and globules had an irregular serrated outline with a dense core which was more obvious after Verhoeff's or Weigert's elastic stain (Fig. 2). The fibres and globules took up the picric acid in the van Gieson technique and the chromotrope in the Masson trichrome stain. Digestion in 0.1 % elastase (Sigma Chemical Co.) in 0.1 M borate buffer at pH 9, showed a progressive loss of elastic fibre staining using the Verhoeff

technique, the initial change being noticed at 15 minutes, with complete loss of characteristic elastic staining at 75 minutes (Figs. 3 and 4) although the fibres still reacted with picric acid. Neither tumour showed evidence of bone infiltration.

ELECTRON MICROSCOPY

The formalin-fixed material was post-fixed for one hour at 4°C in 1 % osmium tetroxide in veronal-acetate buffer (pH 7.4) containing 0.01 mg sucrose per ml and processed to Epon. Blocks of fixed tissue were digested for five hours in 0.1 % elastase in 0.1 M borate buffer at pH 9 and comparable control blocks in buffer alone for five hours before processing to Epon. Ultrathin sections were cut on a Porter Blum MT-2 ultramicrotome and mounted on copper grids. They were then stained with lead citrate and uranyl acetate and examined in a Siemens Elmiskop I electron microscope at 60 Kv.

FIG. 7.

FIG. 7. *Electron micrograph. Elastofibroma. Elastic fibre with a central dense homogeneous core surrounded by dense fibres lacking periodicity. Some of the adjacent collagen fibres show increased density. Compare with Figure 2. $\times 15,000$.*

FIG. 8.

FIG. 8. *Electron micrograph. Elastofibroma. After five hours' digestion in elastase, a residue of the elastic fibres contains fine fibrils and granular material. Collagen fibres around and within the residue retain their characteristic periodicity. $\times 23,000$.*

Tissue from the carotid artery of a 1-year-old child and from the facial skin of an albino adult was similarly processed for comparison of elastic tissue. The fibres and globules of the elastofibroma were clearly recognized as electron dense masses of average diameter 3 to 6 μ (Fig. 5). They contrasted with the much less dense and characteristic collagen fibres amongst which they lay (Fig. 6). In some areas corresponding with the serrated fibres seen in the light microscope, the elastic masses consisted of a homogeneous, extremely dense core surrounded by dense, irregular fibres of diameter 350 Å to 400 Å with an indistinct outline (Fig. 7). These fibres showed no periodicity. At the periphery of the masses it was sometimes difficult to distinguish collagen fibres which were heavily stained and in which

periodicity was ill-defined from the electron dense 'elastic' fibrillary masses. Between the dense, unbanded fibres, there was granular and sometimes dense amorphous material, scanty in some areas but obscuring the fibrillary pattern in others. Transitions from fibrillary to amorphous patterns could be traced. At the periphery of the dense masses, fine fibrils, 80 Å to 100 Å diameter were found, corresponding with fibrils of young collagen.

Elastase digestion produced striking changes in the dense areas although the collagen fibres appeared unaffected and retained their characteristic banding. The electron density of the coarse masses disappeared leaving a delicate granular background with fine fibrils comparable with the young collagen fibrils seen at the edge of the elastic masses (Fig. 8).

FIG. 9.

FIG. 9. *Electron micrograph. Skin. The granular material of the 'elastotic degeneration' extends between collagen fibres showing characteristic periodicity. $\times 60,000$.*

FIG. 10.

FIG. 10. *Electron micrograph. Skin. After five hours' elastase the collagen fibres retain their periodicity. The 'elastotic degeneration' shows a residue of granular and indistinct coarse fibres. $\times 60,000$.*

FIG. 11.

FIG. 11. *Electron micrograph. Artery. A characteristic dense elastic lamina with adjacent collagen fibres. $\times 30,000$.*

FIG. 12.

FIG. 12. *Electron micrograph. Artery. After five hours' elastase digestion the dense elastic lamina cannot be seen although collagen is preserved. $\times 20,000$.*

Isolated, mature, apparently normal collagen fibres could be seen within the areas previously obscured by the dense material.

The sections of solar degeneration of the skin also showed granular and amorphous material lying between characteristic collagen fibres (Fig. 9). However, this granular material for the most part was not so dense as the elastic masses of the elastofibroma, it did not occur in such well demarcated fibres and globules, and the blurred, dense fibrillary pattern seen in many of the elastofibroma masses was not apparent. It was not possible to trace transitions from collagen to the amorphous interfibrillary material in the solar degeneration. In some areas after elastase digestion all the interfibrillary material was lost, but elsewhere granular material remained, giving a pattern of coarse, ill-defined fibres with irregular borders and measuring 30 to 180 m μ diameter (Fig. 10). No normal collagen fibres or fine fibrils were apparent within the elastic residue as had been the case with the elastofibroma after elastase digestion.

The infant artery showed characteristic amorphous elastic fibres with an irregular border measuring 0.8 to 1.6 μ diameter (Fig. 11). Scanty electron-dense particles were embedded in the elastin. Surrounding the elastic fibres there were fine fibrils, apparently young collagen, 80 Å to 100 Å diameter, and mature collagen fibres, 400 Å to 500 Å diameter, with characteristic periodicity. After elastase digestion, only the electron-dense particles remained in the elastic fibre (Fig. 12), a pattern quite unlike that seen in the elastofibroma or in the solar degeneration of the skin. In the light microscope sections, the arterial elastic lamina did not stain either with Verhoeff's elastic stain or with picric acid in the van Geison technique after digestion in elastase.

DISCUSSION

Although frequently misdiagnosed clinically as sarcoma, elastofibroma neither behaves like, nor has the morphological appearances of, an aggressive neoplasm. It has features which clearly distinguish it from other fibromatoses. Much of the discussion on the previously reported cases has been based on the cause of the lesion and the nature of the 'elastic' fibres found there in. Stemmermann and Stout (1962) suggested that the elastic fibres and globules had a core of elastin which was removed by digestion in elastase, and this core was surrounded by reticulin fibres resistant to elastase but also impregnated with a material which stained for elastin. Electron microscopy enables the structure of these masses to be studied in more detail. They show that apparently normal collagen fibres and fine fibrils are to be

found embedded in the granular elastic masses and that the fibres resist digestion with elastase (Fig. 8). However, the amount of collagen found after digestion would not account for the material stained with picric acid after elastase as seen in the light microscope. In addition to the collagen fibres, many of the elastic fibres and globules have a core which consists of extremely dense amorphous material resembling true elastin rather than collagen (Fig. 7). The material which surrounds the dense core is of much interest for it is clearly fibrillary and superficially resembles collagen. However, it differs from collagen in its lack of periodicity and in the blurred outline of the fibres. The adjacent collagen fibres frequently show some increased density and partial loss of definition of the characteristic 640 Å banding. It would seem reasonable, therefore, to suggest that the peripheral fibrillary areas of the elastic masses represent denatured collagen and that these may become converted into the homogeneous dense core. This is very much in agreement with the findings of Stemmermann and Stout (1962), the fibrillary material that they demonstrated corresponding with the dense, blurred fibres seen in the electron microscope. It also supports the work of Keech, Reed, and Wood (1956) and Keech and Reed (1957) who suggested that collagen and elastin are not two separate and distinct entities but are probably associated *in vivo*, intermediate forms being found. This transition may also account for the variable staining of arterial elastic fibres reported by Rodgers, Puchtler, and Gropp (1967). No direct transition could be traced between the granular elastin and recognizable collagen although the two were frequently found in close association. Any transition apparently occurs through the dense, non-banded fibres forming the periphery of some elastic fibres and globules. The reaction to elastase showed differences between the elastic fibres of arterial wall and the elastin of elastofibroma and cutaneous solar degeneration. In the light microscope sections of the elastofibroma, residual material could be stained with picric acid after elastase digestion whereas no stainable material could be demonstrated at the site of the elastic fibres in the arterial wall. These observations were confirmed in the electron microscope where residual granular and finely fibrillary material was found only in the elastofibroma globules after elastase digestion, even where this was prolonged in excess of that required to inhibit elastic staining seen at light microscope level. Elastotic degeneration of skin has some similarities to the elastic fibres and globules of the elastofibroma (Stemmermann and Stout, 1962). In the electron microscope there is close similarity between the granular material impregnating collagen fibres but

after elastase digestion the skin pattern was variable and where residual material was to be found this was unlike that remaining in the elastofibroma and assumed a coarse, ill-defined fibrillary pattern. The characteristic fibres and globules of the elastofibroma were not seen in the solar skin degeneration.

Neither of these cases showed evidence of pseudoxanthoma elasticum nor was there evidence of calcification of any of the elastic masses. Calcification was a prominent feature of the ultrastructure of the abnormal fibres of pseudoxanthoma elasticum studied by Huang, Steele, Kumar, and Parker (1967).

From the histochemical and electron microscopic features it appears that the characteristic fibres and globules of the elastofibroma arise by denaturation of collagen fibres and accompanying impregnation by material that reacts as for elastin. However, there are distinctive morphological differences between this material and the elastin of arterial wall elastic laminae, elastotic degeneration of skin, and the

abnormal elastic fibres of pseudoxanthoma elasticum. The cause of this change is not apparent but the theory of friction proposed by Jarvi and Saxén (1961) explains the situation of origin of the lesions. From the morphology and clinical behaviour there is little to suggest that elastofibroma is a true neoplasm but rather that it is a reactive fibromatosis.

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