RECURRING DIGITAL FIBROMA OF INFANCY

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Most tumours of the hand are clearly defined with regard to their origin, course and prognosis, but a few remain with these aspects still obscure. An example is the recurring digital fibroma of infancy.

CASE REPORTS

Case 1—A boy aged six months was seen in 1969 because of a swelling on the back of the distal part of the right middle finger. He was the fourth child of healthy parents. The tumour had appeared in the third month of life; it was about 5 millimetres in diameter, firm, not tender, and fixed to the skin but not to the underlying tissues. Biopsy showed a dermatofibroma with no sign of malignancy. The tumour then started to grow more rapidly, and it was decided to excise it completely and apply a skin graft. Three months later a recurrence was excised. After three more months a second recurrence had occurred and the parents refused further operations. Now after four years this has disappeared and only a small local thickening is evident on the lateral aspect of the finger where the graft meets normal skin.

Case 2—A girl aged four months was first seen in 1970 with swellings on the distal parts of the middle and ring fingers of both hands (Fig. 1); according to the mother they had appeared after birth and were still growing. This girl also had pigmented marks over the left temporal region and an abundance of the vermillion of the lower lip. The tumours were of firm consistency and more or less encircled the fingers. Both ring fingers showed a marked flexion contracture of the distal joint.

The tumours were excised and the defects covered with skin grafts. At operation it was clear that this could not be a radical excision. The histological diagnosis was dermatofibroma,

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with no indication of malignancy. Recurrences soon presented and by two months the
situation was worse than ever, especially on the left hand (Fig. 2). It was decided to wait and
see. At four months the tumours were still growing (Figs. 3 and 4), but at eight months only
minor further growth was noted and by fourteen months growth seemed to have stopped.

FIG. 2
Case 2—A photograph of the left hand only one
month after excision of the tumours.

FIG. 3
Case 2—The hands four months after operation.

FIG. 4

Since then gradual regression has been observed (Fig. 5), though in 1973 two new tumours
appeared, on the left index and little fingers (Fig. 6). The original tumours are now much
reduced in size and the flexion contractures have not changed. The left third finger shows
retarded growth, and a radiograph of the left hand shows deformity of the third and fourth
metacarpals (Fig. 7).
Case 3—A girl aged four months was seen in 1971 because of swellings on the distal parts of all the fingers and of one toe, first noticed during the first month of life. She was the first child of healthy parents and there had been no complications during the pregnancy. The child had a patchy brownish discoloration of the forehead, a pre-auricular skin tag, a minor degree of hypertelorism and a local abundance of the vermilion of the lower lip.

Most of the tumours were only a few millimetres across, but two encircled the fingers. They were firm, not tender, and fixed to the skin; the larger ones were fixed to the deep tissues. Radiologically the third and fourth metacarpals of the right hand were shortened, as were the second, third and fourth metatarsals of the right foot. The right fourth toe was deviated dorsally and displayed a tumour. Biopsies showed all the tumours to be dermatofibromas, with no signs of malignancy.

It was decided to excise all the swellings and where necessary to apply skin grafts. This
was done, but the deformed toe was amputated. One month after these operations it was clear that the tumours were recurring. It was then decided to wait and see. The tumours now grew more slowly; over the next year there was some increase in size but also an occasional suggestion of regression. Two new tumours developed, on a toe of each foot, but at the last review in January 1974 the condition was essentially unchanged.

**FIG. 7**
Case 2—A radiograph of both hands at the age of 3½ years, showing various abnormalities of the metacarpals and flexion contractures of three fingers.

**FIG. 8**
Case 2—To show the histological picture of recurring digital fibroma of infancy. (Haematoxylin and eosin, × 400.)

**HISTORICAL ASPECTS**

In 1965 Reye reported six cases of digital fibrous tumours in children. He claimed that the lesions differed from other forms of fibromatosis in three respects—clinically by being limited to the fingers and toes of infants and by having a remarkable tendency to recur, and...
morphologically by the presence of cytoplasmic inclusion bodies. Earlier reports of these tumours under different names make no mention of the cytoplasmic inclusion bodies, which are hard to recognise by routine staining. Reye also thought of a viral causative agent, but having no more unfixed tissue he could make no attempt to isolate a virus.

Judging from the clinical and pathological description, there is no doubt in our opinion that Jensen, Martin and Longino (1957) were reporting true cases. They concluded that this was a malignant tumour of multicentric origin and probably of an infective (viral) etiology. As none of their cases metastasised and as mitotic figures were infrequent in their histology, they must have been following the criteria for malignancy of Posch (1956), who regarded local recurrence within a year as a sign of malignancy.

Shapiro (1969) reviewed the literature and found twenty-one acceptable cases. The lesions were equally divided between boys and girls. They were mostly on the fingers and were multiple in twelve cases. The tumours were either congenital or appeared in the first few months of life, except in three cases. In no case was metastasis recorded. Burry, Kerr and Pope (1970) and Battifora and Hines (1971) studied these tumours by electron microscopy and tissue cultures. Neither team was able to isolate a virus. Grunnet, Genner, Mogensen and Myhre-Jensen (1973) collected thirty published cases and added one case with a follow-up of ten years.

**PATHOLOGY**

Macroscopically the tumours begin as small nodules which sometimes grow slowly, sometimes rapidly, and may reach a great size. They are frequently multiple. The common location is on the lateral surfaces of the distal parts of the fingers and toes, less frequently on the dorsal aspects. The adjoining sides of fingers are often involved. The overlying skin is normal in colour or slightly reddened. On palpation the tumours are firm in consistency, fixed to the skin and sometimes to the underlying tissues. On section they are not encapsulated, are greyish-white in colour and show a fibrous structure.

Microscopically the epidermis is not involved. The dermis is replaced by tumour consisting of interlacing bands of fibrous connective tissue and an abundance of collagen. The nuclei vary in size; mitotic figures are infrequent (Fig. 8).

By electron microscopy two forms of fibroblast can be distinguished, one with a large lobulated nucleus and scanty endoplasmic reticulum, the other with a small flattened nucleus and more endoplasmic reticulum. These cells and the collagen are entirely normal. Ahlqvist, Pohjanpelto, Hjelt and Hurme (1967), Shapiro (1969), Burry et al. (1970), Battifora and Hines (1971) and Grunnet et al. (1973) refer to the cytoplasmic inclusion bodies that were first described by Reye (1965). Apparently they are very hard to find in routine stainings with haematoxylin and eosin. With phosphotungstic acid haematoxylin they stain deep purple and they give a negative periodic acid-Schiff reaction (Shapiro 1969). Most authors state that the inclusion bodies are pyroninphyllic but in Ahlqvist's case (1967) they were not.

In our three cases we were unable to find cytoplasmic inclusion bodies despite a thorough search. Otherwise the histology was identical with the descriptions given in the literature.

**Virology**—Most authors believe a virus to be the causative agent but none has been able to isolate a strain. Only Pohjanpelto and his colleagues (1967) found a cell-transforming agent in the filtrate which caused clustering of fibroblasts in a normal fibroblast culture. We too have been unable to demonstrate a virus, despite having enough unfixed material and excellent tissue cultures.

**TREATMENT**

Believing in the malignancy of this tumour, Jensen et al. (1957) stated that wide excision and skin grafting should be the preferred treatment, with amputation for advanced cases. One of their cases was treated with x-rays but without apparent effect. Shapiro recommended conservative surgical removal because no case of metastasis had been reported. Grunnet et al.
advocated the widest possible excision without impairing the function of the part. In adopting this conservative attitude they referred to Stout (1954) who followed a boy with an untreated tumour for ten years; the tumour persisted but no metastasis occurred. In our own cases we first thought that complete excision with skin grafting was the treatment of choice. However, as this is apparently a disease of childhood and never seen thereafter, it may be that the tumours disappear spontaneously. If the tumour is indeed caused by a virus, the anti-virus titre might in due time rise to such a level that the virus is killed and the tumour dissolved.

We are left with several questions. Firstly, are cytoplasmic inclusion bodies a prerequisite for the diagnosis? Secondly, are the inclusion bodies structures related to the action of viruses or are they aggregates of fibrillary proteins, products of cell degeneration? Thirdly, if the tumour is indeed caused by a virus, could it be cured by vaccination? Fourthly, will the tumour disappear spontaneously by some immunological mechanism or is surgery the treatment of choice? In the literature we have found two cases with a long-term follow-up (Stout 1954, Grunnet et al. 1973). Two of our own cases have now been followed for three years and one for four years, and in all three we have observed definite spontaneous regression. We therefore believe a strictly conservative approach to be the right one, realising that this calls for a high level of confidence between parent and surgeon. And finally, are the congenital deformities described in our Cases 2 and 3 part of the syndrome?

SUMMARY

1. Three typical cases of recurring digital fibroma of infancy are reported, with a follow-up of three to four years. In each case excision of the tumours during the first year of life was followed by recurrence and then by some degree of slow spontaneous resolution, in one case complete.

2. A strictly conservative approach is recommended for three reasons: the difficulty of complete excision, the tendency to spontaneous regression and the facts that no case of metastasis and no case of persistence into adult life have yet been reported.

3. Cytoplasmic inclusion bodies could not be demonstrated in the biopsy material from these cases, nor any virus.

REFERENCES


