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A CONSECUTIVE 7-YEAR SERIES OF 1331 BENIGN SOFT TISSUE TUMOURS

Clinicopathologic Data. Comparison with Sarcomas

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A consecutive 7-year series of 1331 benign soft tissue tumours was analysed and compared with the data of 72 sarcomas diagnosed in the same period (April 1970 to April 1977). Lipoma was the most frequent benign tumour entity, accounting for almost half of the entire series. Different entities had different male to female ratios and preferred locations and, furthermore, differed from sarcomas in this regard. The factors which discriminated most in the clinical differential diagnosis benign vs. malignant were tumour size and location.

At the first visit to the general practitioner or surgeon it is recommended that patients with large (≥50 mm) and/or deep seated tumours (beneath or fixed to superficial fasciae) or tumours obviously malignant for other reasons (invasion of nerves, vessels or bone) should be transferred, leaving their tumours untouched, to a centre for soft tissue sarcomas for further diagnosis and treatment.

Key words: benign soft tissue tumour; first visit; sarcoma

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Previous work on benign soft tissue tumours has dealt mostly with separate histiologic entities and their variants. Detailed surveys on the histopathology of fibroblastic-histiocytic, neurogenic, and vascular tumours (Mackenzie 1970, Abell et al. 1970, Johnson 1976) have recently been published. A good deal of attention, has also been paid to the so-called pseudosarcomatous lesions (Dahl & Angervall 1977). Only a few publications devoted to unselected series of benign soft tissue tumours, however, have appeared (Stout 1953, De Rubertis & Lettieri 1969, Templeton 1973). Therefore, in the following a consecutive series of 1331 benign soft tissue tumours is presented and in some respects compared with sarcomas.

MATERIAL AND METHODS

The material comprises all benign soft tissue tumours seen at the Surgical Pathology Laboratories at our institute during the 7-year period April 1970 to April 1977 (WHO code no. 197 connective tissue, 190–191 skin and integumentary system, 193, 1–9 peripheral nerves). New cases of soft tissue sarcomas diagnosed in the same period were used for comparison. Material sent for consultation, not primarily examined in our laboratories, was omitted. Tumour specimens were examined primarily by the author or microscopical slides were reviewed. Diagnoses were registered according to the WHO classification of soft tissue tumours (WHO International Histological Classification of Tumours). Clinical information on individual cases was obtained from microscopy request sheets and/or clinical records. Tissue specimens were formalin fixed, paraffin embedded and sectioned at 6–8 μm. The sections were routinely stained with haematoxylin-eosin and picro-fuchsin-haematoxylin (van Gieson) and occasionally with periodic acid-Schiff (P.A.S.), for reticulin (Gordon & Sweet), elastin and haemosiderin (Perls).

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RESULTS

During the 7-year period, April 1970 to April 1977, a total of 1331 benign soft tissue tumours were detected. In the same period 72 new patients with malignant tumours of the somatic soft tissues were seen (29 malignant fibrohistiocytomas, 13 fibrosarcomas, 10 liposarcomas, 5 leiomyosarcomas, 5 embryonal and/or alveolar rhabdomyosarcomas, 4 extraskeletal chondro- and osteosarcomas, 3 epithelioid and synovial sarcomas, 1 dermatofibrosarcoma protuberans, 2 unclassified sarcomas), giving a benign to malignant ratio of 18.5. In the same period the total number of requests for histopathological examinations was 61,557. Thus the entire soft tissue tumour material accounts for only about 2 per cent of the total load of the surgical pathology laboratories.

Table 1. A consecutive series of 1331 benign tumours of the somatic soft tissues (1970–77)

Lipoma		640	48.1%
Fibro-histiocytic tumours		244	18.3%
Dermatofibroma	211		
Benign giant-cell synovioma	32*		
Atypical fibroxanthoma	1		
Vascular tumours		181	13.6%
Haemangioma	157		
Lymphangioma	19		
Glomus tumour	4		
Haemangiopericytoma	1		
Fibroma, fibromatosis		140	10.5%
Cutaneous fibroma	80		
Elastofibroma	2		
Fibromatosis palmaris & plantaris	43		
Fibromatosis abdominis (desmoids)	5		
Fibromatosis colli (torticollis)	1		
Nodular fasciitis	2		
Others	7		
Neurofibroma, neurilemmoma		68	5.1%
Leiomyoma, angiomyoma		51	3.8%
Granular cell tumour		4	0.3%
Myxoma		3	0.2%

* 229 ganglia and bursa cysts and 15 villonodular (pigmented) synovitides diagnosed in the same period are not included among the tumours.

Table 2. Age and sex distribution of 1331 patients with benign soft tissue tumours

	Age range (years) (90%)	Sex male/female ratio
Lipoma	20–69	0.9
Dermatofibroma	20–69	0.5
Benign giant-cell synovioma	20–69	0.8
Angioma (haem.+lymph.)	0–59*	0.7
Cutaneous fibroma (molle+durum)	20–69	1.0
Palmar & plantar fibromatosis	20–69	3.0
Neurofibroma, neurilemmoma	20–69	0.9
Leiomyoma, angiomyoma	10–79	0.8

* 40% of patients aged below 20 years.

The histologic entities of the entire series, and the distribution of common entities as regards age, sex and anatomical region are shown in Tables 1, 2, 3. Benign tumours showed a moderate female preponderance, except for palmar and plantar fibromatoses. By comparison, the male to female ratio in the 72 sarcomas was 2.0. The age range in the benign series (comprising more than 90 per cent) was 20 to 69 years, except in vascular tumours and, to some extent, smooth muscle tumours; in the malignant series it was 20 to 79 years with the exception of the group of embryonal and alveolar rhabdomyosarcomas, where 4 out of 5 patients were below 10 years of age.

Lipoma was the most frequent entity, making up almost half of all cases of benign soft tissue tumours (48.1 per cent). Male to female ratio was 0.9. Forty-four patients (6.9 per cent) had more than one tumour; in all 707 lipomas were examined. Less than 1 per cent of lipomas were deep seated, three were intramuscular and two intraarticular (lipoma arborescens). Range of tumour size (comprising more than 90 per cent) was 10–50 mm (maximum 200 mm). Usually the tumours were grossly lipomatous in character.

Fibrohistiocytic tumours were next in frequency, accounting for 18.3 per cent of cases.

Table 3. Distribution (per cent) of benign soft tissue tumours according to anatomical regions

	Hands (+ wrists)	Forearms (+ elbows)	Arms (+ shoulders)	Head & neck	Trunk (+ mammae)	Thigh (+ hips & groins)	Legs (+ knees)	Feet (+ ankles)
Lipoma	1.6	9.7	14.1	12.7	43.4	13.0	5.0	0.5
Dermatofibroma	8.0	5.7	14.6	3.8	13.2	20.3	31.6	2.8
Benign giant-cell synovioma	74.2	3.2	0	0	0	6.5	6.5	9.6
Haemangioma	22.2	5.9	8.9	17.8	20.7	8.9	8.9	6.7
Lymphangioma*								
Cutaneous fibroma	7.0	4.2	2.8	8.5	43.7	31.0	1.4	1.4
Fibromatosis**								
Neurofibroma, neurilemmoma	16.0	9.3	12.0	13.3	18.7	8.0	12.0	10.7
Leiomyoma, angiomyoma	16.7	8.3	4.2	0	2.1	10.4	47.9	10.4

* 7 of 19 lymphangiomas were located in the head & neck.
** Of 54 fibromatoses, 29 were palmar, 14 plantar, 5 abdominal (desmoids).

The dermatofibroma (syn.: xanthofibroma, sclerosing angioma) was the benign tumour with the greatest female preponderance. More than 50 per cent of the tumours were located in the lower extremity. Two occurred in previous tattoos. Dermatofibromas, generally less than 30 mm in largest diameter, were white with yellow or brownish areas and sometimes causing a characteristic bluish colour of the skin. The benign giant-cell synovioma (syn.: xanthomatous, giant-cell tumour of tendon sheaths) was located almost exclusively in the hands (and wrists). The tumours were typically lobulated, yellow or yellowish brown due to the content of lipids and haemosiderin, and had diameters below 30 mm. A further 15 cases of non-localised, villonodular synovitis, mostly in the knee joints, were not registered as tumours. Neither were 133 ganglia and 96 bursa cysts diagnosed in the same 7-year period.

Vascular tumours rated third in frequency, the bulk of this group being haemangiomas (77 cavernous, 48 capillary, 9 arteriovenous, 5 pyogenic granuloma, 5 benign haemangioendothelioma, 14 unclassified) and lymphangiomas (14 cavernous, 5 capillary or unclassified). Almost half of the number of lymphangiomas were "cystic hygromas" of the neck. Forty per cent of the patients with vascular tumours were aged below 20 years. Furthermore, it is noteworthy that 20 per cent of the vascular tumours were located in the hands (and wrists). Hands, trunk and head and neck region contained 60 per cent of all vascular tumours. This group comprised also four patients with glomus tumours (3 male, 1 female, aged 40–75 years), three located in the hand and arm, one in the cheek, and one haemangiopericytoma, occurring in a 77-year-old woman subcutaneously in the frontal region. Haemangiomas were grossly dark blue or reddish and usually below 30 mm in diameter. Lymphangiomas were grey-white and often cystic, measuring up to 60 mm in diameter.

Fibromatous tumours accounted for 10.5 per cent of all benign soft tissue tumours. Cutaneous fibromas appeared either as pedunculate, often fat–containing tumours (fibroma molle) or as sessile, hard tumours (fibroma durum). Most of them were fairly small, only exceptionally up to 50 or 60 mm in diameter. Only two elastofibromas were diagnosed, both located in the back of elderly men. Of the fibromatoses, 29 cases were palmar, 14 plantar and 5 abdominal desmoids. Furthermore, a 1½-year-old boy had

sternocleidal fibromatosis, and two men (aged 21 and 23 years) had nodular fasciitis of the forearm and the back. Other fibromatoses included four patients with keloids (3 males, 1 female), two girls (4 and 6 years old) with juvenile aponeurotic fibromas, and one girl (17 years) with "knuckle-pads". Abdominal fibromatosis (desmoids) occurred only in women. Palmar and plantar fibromatosis mostly measured from 10 to 30 mm, and were often whitish with faintly yellow streaks centrally. Abdominal desmoids measured up to 70 mm in largest diameter.

Neurofibroma, neurilemmoma were diagnosed in 68 patients; six had more than one tumour and four patients had neurofibromatosis Recklinghausen. None of them developed malignant Schwannomas within the period of observation. Generally, it was difficult to make a precise distinction between neurofibroma and neurilemmoma; 27 of the cases, however, were considered pure lemmomas. The neurofibromas/neurilemmomas were fairly evenly distributed in the anatomical regions, showing a slight female preponderance, and were usually small and sometimes gelatinous on sectioning.

Leiomyoma, angiomyoma (cutaneous, subcutaneous) occurred over a wide age range and had a moderate female preponderance. Almost 50 per cent of all smooth muscle tumours were located in the legs (and knee regions). In those patients, for whom the clinical records contained information as to patient symptoms, one fourth had complained of pain and tenderness referable to these tumours. No difference between leiomyoma and angiomyoma (vascular leiomyoma) as regards age and sex of the patients, location and symptoms were noted. The leiomatous tumours were usually small, in a few cases up to 40 mm and only exceptionally 70 mm in diameter, hard in consistency, sometimes like cartilage and grey-white with darker or yellowish areas and hyalinous on sectioning.

Granular cell tumour was diagnosed in four patients, two in the hands related to traumatic neuromas (21-year-old male and 21-year-old female) and two subcutaneously in the mammary regions (52- and 54-year-old females). These tumours were below 10 mm in diameter and whitish. *Myxomas* were diagnosed in three patients (two males 74 and 71 years old and one female 26 years old). All three tumours were grossly gelatinous, but well-demarcated and encapsulated. Two were intramuscular and rather large, i.e. more than 50 mm in diameter.

Clinical data in benign soft tissue tumours are usually very sparse. Generally, the history stated that an intumescence had been present for some time, mostly several months or years. Tenderness and pain were noted in cases of neurogenic and leiomyomatous tumours, most frequently in the latter (25 per cent). One dermatofibroma, three haemangiomas and two benign giant-cell synoviomas recurred out of the entire series of benign soft tissue tumours, obviously because of incomplete primary removal. Multiple recurrences were not encountered, and none of the patients in the benign series have died because of the benign soft tissue tumours or complications from operation.

Relation benign-to-malignant as regards tumour size and location. Most benign soft tissue tumours were small; only 50 lipomas, 2 lymphangiomas, 3 neurofibromas/neurilemmomas, 1 dermatofibroma, 2 cutaneous fibromas, 2 abdominal fibromatoses (desmoids) and 2 myxomas, i.e., almost 5 per cent of all benign soft tissue tumours, measured 50 mm or more in diameter. By contrast, more than 50 per cent all malignant soft tissue sarcomas were large, i.e. $\geq$50 mm in diameter.

With the exception of benign giant-cell synoviomas and fibromatoses, almost all benign soft tissue tumours were superficially located, i.e. in the skin, in subcutaneous fat and sometimes including the superficial fasciae. Out of 72 patients with malignant soft tissue tumours (including 1 case of dermatofibrosarcoma protuberans) 19 tumours were located in the skin, subcutis and superficial fasciae and of these only 9 tumours were entirely superficially located and freely movable in relation to the underlying fasciae; the rest of the sarcomas were deep seated (73 per cent). Less than 1 per cent of lipomas were deep seated (intramuscular, intraarticular). A few haemangiomas and lymphangiomas had, in addition to a superficial location, deep extensions, too. Only a few of the neurogenic tumours

and none of the leiomyomatous were deep seated.

Benign giant-cell synoviomas occurred almost exclusively in the hands (fingers) and wrists, usually grossly related to tendons or tendon sheaths. Fibromatoses were primarily situated in muscle aponeuroses, mostly palmar and plantar; in the abdominal fibromatoses (desmoids) there was generally extensive muscle infiltration, as well. Dermatofibromas and leiomyomas/angiomyomas were always superficially located, mostly in the legs. In the malignant soft tissue tumours the thigh (and buttocks) was the preferred anatomic location (30 per cent).

DISCUSSION

In a series of 7,337 benign soft tissue tumours, of which 6,364 would be registered according to our classification, Stout (1953) found 37.9 per cent lipomas, 31 per cent fibromas (including dermatofibroma) and fibromatosis, 19.4 per cent angiomas, 5.1 per cent benign synoviomas and 3.4 per cent leiomyomas. Other tumour entities, such as granular cell tumour, benign mesenchymoma, and rhabdomyoma, together accounted for less than 3 per cent. Neurogenic tumours were not included. In Templeton's material of soft tissue tumours in Uganda (1973), 997 were benign: 26.4 per cent lipomas, 26.1 per cent fibromas and fibromatoses, 18.0 per cent vascular tumours (13.1 per cent haemangiomas, 4.2 per cent lymphangiomas, 0.7 per cent glomus tumours), 14.4 per cent neurogenic tumours, 10.4 per cent leiomyomas, and 3.7 per cent other categories. Taking into account the geographical differences, our figures compare surprisingly well with these two series, with the exception of a greater frequency of lipomas in our series.

In 352 lipomas Adair et al. (1932) found 43 per cent located in the trunk, and 6.7 per cent of patients had multiple lipomas. The age distribution was also the same as in our material, but their male to female ratio was lower (0.4). In Geschickter & Lewis' material (1935) of (fibrous) connective tissue tumours from the Surgical Laboratory of Johns Hopkins Hospital there were 8 dermatofibromas, 125 keloids, and 20 desmoids. Both dermatofibroma and keloid tumours were related to previous trauma. In our series, in which keloid was comparatively rare probably because of the different racial background, relation to previous trauma could not be analysed but two dermatofibromas occurred in tattoos. Groos & Wolbach (1943) found sclerosing haemangiomas (syn.: histiocytoma, dermatofibroma) located in the lower extremity in about 50 per cent and Kauffman & Stout (1961) also found histiocytic tumours, in this case in children, to be most common in the lower extremity. Our own figure for the lower extremity is 55 per cent Geschickter & Keasby (1935) found that 43.7 per cent of peripheral (vs. visceral) angiomas occurred in patients up to the age of 20 years, a figure similar to ours. Boman (1939), like us, found most haemangiomas (63 per cent) to be located in the head and neck, trunk and hand. In our series peripheral neurogenic tumours were less frequent than in other materials (Templeton 1973), and the neurofibroma to neurilemmoma ratio was lower. A clear distinction between neurofibroma and neurilemmoma, however, was not possible, as was the case in Saxén's material (1948). Solitary cutaneous and subcutaneous leiomyomas were found especially on the extensor surfaces of the lower extremity in about half of all cases in Stout's material (1937), with a slight preponderance in females and making their appearance at any age. The characteristic pain and tenderness of this tumour was stressed, and the opinion was expressed that leiomyomas together with glomus tumours form the bulk of the painful subcutaneous tumours (tubercula dolorosa) described by earlier authors. Christopherson (1950) and Fisher & Helwig (1963) reported a further five and four cases, respectively, of solitary (non-genital) leiomyomas, the angiomyomas not being included. In our own series, 30 out of 51 benign smooth muscle tumours were angiomyomas (vascular leiomyomas). As there were no differences between leiomyomas and angiomyomas as to clinical and pathological findings they were grouped together. In our series benign giant-cell synovioma (syn.: xanthomatous, giant-cell tumour of tendon sheath, localised nodular synovitis) (Lichtenstein 1955) occurred most fre-

quently in the hands (and wrists). In Wright's series (1951) 86 per cent were situated in the same topographical region and particularly in the digits.

Whereas the relative frequency of individual tumour entities in large unselected series of benign soft tissue tumours seems fairly constant as does the clinicopathologic data, the ratio benign/malignant is largely determined by the degree of centralization in the diagnosis (and treatment) of soft tissue sarcomas. Therefore, Stout's figure (Stout & Lattes 1967) is much lower than ours (5.5 vs. 18.5).

The age distribution in benign vs. malignant soft tissue tumours does not show much difference; the appearance of most tumours is spread over a wide age range of adult life. However the female preponderance in the benign group, with great differences between individual entities, is reversed to a strong male preponderance in the malignant group (2/1). As to anatomic location each benign entity has its characteristic location and, in contrast to the malignant tumours, no preference for the thigh.

Benign soft tissue tumours are usually small, only 62 out of the 1331 benign soft tissue tumours (5 per cent) measured 50 mm or more in diameter, whereas 38 out of 72 sarcomas (53 per cent) had diameters ≥50 mm. Except for certain tumour entities such as fibromatoses and benign giant-cell synovioma, benign tumours of the somatic soft tissue are located superficially. All in all, in our material only about 10 benign tumours, i.e. less than 1 per cent, were deep seated. In contrast, almost three quarters (73 per cent) of all malignant tumours of the somatic soft tissue were deep seated, i.e. beneath the superficial fasciae. Only 12.5 per cent of the cases with malignant soft tissue tumours were located superficially and freely movable in relation to the underlying fasciae.

The outlook for patients with soft tissue sarcomas is mainly determined by two factors: 1) the nature of the tumour, i.e. histogenetic type, grade of malignancy and stage of disease 2) the primary treatment, i.e. surgery and radiotherapy and/or chemotherapy (Suit & Russell 1977, Russell et al. 1977). As far as possible primary treatment should be instituted in a centre for soft tissue sarcoma. Not every patient with a soft tissue tumour, however, can be transferred to a special centre. In the light of our experience it is suggested that any patient with a large (measuring about or more than 50 mm in diameter) and/or deep seated soft tissue tumour, as well as a tumour obviously malignant for other reasons (invasion of nerves, vessels or bone), should at the primary visit to the general practitioner or surgeon be transferred, leaving the tumour untouched, to a centre for soft tissue sarcoma for further diagnosis and treatment.

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