# Concomitant Right Subscapular and Left Olecranon Elastofibroma Followed by Inversion of the Lesions: Case Report

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**Abstract.** *Elastofibroma is a benign, poorly circumscribed, tumor-like condition involving, in the vast majority of cases, the subscapular region of elderly individuals, though isolated cases have been seen in the deltoid muscle, infraolecranon area, hip, thigh and stomach. It is characterized by accumulated abnormal elastic fibres and is generally regarded as a reactive process, an unusual fibroblastic pseudotumor. Multiple elastofibromas have been reported to occur in the scapula and olecranon and in the scapula and ischium, whereas literature reports of multiple elastofibromas in the same patient are rare. The case of concomitant, asynchronous double elastofibroma in the same patient is described. A 69-year-old woman presented with right subscapular and left olecranon swelling associated with pain and a clicking sensation during certain arm movements. Some months later the patient developed asymptomatic left subscapular and right olecranon swelling. All the lesions, which were subsequently disgnosed as elastofibromas, were removed.*

Elastofibroma is a benign, poorly circumscribed, tumor-like condition involving, in the vast majority of cases, the subscapular region of elderly individuals, though isolated cases have been seen in the deltoid muscle, infraolecranon area, hip, thigh and stomach. It is characterized by accumulated abnormal elastic fibres and is generally regarded as a reactive process that results in an unusual fibroblastic pseudotumor (1). Jarvi and Saxon first described the tumor in 1961 (2). Since then, other authors have documented the gross and histopathological features of this benign tumor, which is well characterized in the literature (3-6).

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Elastofibromas are usually found in active individuals beyond their 50th year (3-6), but may also affect children (7). In elderly patients, this tumor was incidentally found in up to 2% by CT imaging (8). Autopsy studies have reported an even higher incidence (13% to 17%), with pre-elastofibroma-like morphological changes being found in as many as 81% of the autopsies (9, 10). Its prevalence in those over 55 years of age is reported to be as high as 24% (10). Most (>50%) elastofibromas are asymptomatic (11). When symptomatic, patients may present an enlarging soft tissue mass combined with shoulder pain, discomfort, "locking" or "snapping" (11).

The fact that multicentre and familial cases have been described points to the existence of a predisposing background. At surgery, the lesions are usually found at the apex of the scapula, beneath the rhomboid and latissimus dorsi muscles. The right side is more often affected than the left, while bilaterality is frequent. A periosteal origin has been suggested.

Although the etiology of this tumor remains unclear, its prevalence is higher in persons who perform manual labor involving the shoulder girdle. Thus, it has been suggested that repeated trauma due to mechanical friction of the scapula against the ribs may induce this pseudotumoral process. Although this theory may also explain the right-sided prevalence of elastofibromas, it should be borne in mind that as many as 66% of cases are bilateral (4, 10, 12). In rare cases, the same individual may be affected by multiple elastofibromas. Moreover, up to one third of elastofibroma patients have a family history of tumors, which supports a genetic, as opposed to a traumatic, origin of such tumors (4). Recognition of the lesion is important as the differential diagnosis includes other tumors that may be benign or malignant.

This report describes the case of a 69-year-old woman who presented with right subscapular and left olecranon swelling associated with pain and a clicking sensation during

certain arm movements. Some months later the patient developed asymptomatic left subscapular and right olecranon swelling.

## Case Report

In February 2007, a 69-year-old housewife was admitted to the Department of Plastic Surgery of the University "La Sapienza", Rome, with a palpable mass located to the right of the subscapular area and another lesion in the left olecranon region; both lesions had been present for 1 year. The patient referred the presence of a non-tender, fixed mass in the right, posterolateral part of her chest wall adjacent to the scapula, joint stiffness, and a mild pain and clicking sensation of the posterolateral part of her chest when she moved her arm (Figure 1). She did not refer any pain due to the olecranon lesion.

The physical examination revealed a swelling measuring about 7×7 cm in the right subscapular region that was evident when the arm was mobilized, as well as a 5×5 cm swelling in the left elbow. The ultrasound examination demonstrated hyperechogenic swellings adhering to the periosteal plane, which led to a presumptive diagnosis of advanced fibrolipoma.

CT imaging of the thorax demonstrated two solid soft tissue masses with well-defined margins and a heterogeneous signal intensity. These lesions were located in the subscapular (Figures 2 and 3) and elbow areas, respectively. Although the neo-formations were compatibile with a common lipoma, surgery was performed under general anesthesia owing to the deep site and considerable size of the lesions.

A retrothoracic surgical approach was used, including a subscapular incision parallel to the 6th-7th ribs on the right side and a longitudinal incision along the olecranon. A marginal excision was excluded because the neo-formations were strongly adherent to the ribs and olecranon. Inelastic, soft, non-capsulated, whitish deformations were found in the subscapular area (size 8×7.5×3 cm) and elbow (size 4×4×3 cm).

The macroscopic investigation revealed that the two lesions were characterized by an irregular, poorly defined fibroelastotic mass, with a slightly rubbery, elastic consistency. The cut surface showed strands of white and yellow tissue caused by the entrapment of fatty remnants, similar to a “checkerboard” pattern. The tumors were not encapsulated.

The histological investigation on both masses revealed that they were composed of fibrous, collagenous strands and plump, occasionally elongated, mostly round-shaped elastic fibres, which were densely packed. The elastic structures typically formed discs or globules and sometimes appeared in an “asbestos-body-like” fashion. The lesions were predominantly hypocellular with fibrocytic and fibroblastic cells without atypia or mitotic activity (Figure 4). The result of this histopathological examination was suggestive of elastofibroma.

An unusual event occurred 5 months later: the patient presented the same symptoms though with the site of the lesions inverted. The physical examination revealed a swelling measuring about 6×6 cm in the left subscapular region that was evident when the arm was mobilized, and another 3×2 cm lesion in the right elbow. The same surgical approach was used to remove these two neo-formations. The result of the histological examination on both masses was again suggestive of elastofibroma. To date, one year after the second surgical procedure, the patient’s local and general conditions are good, with no signs of lesion recurrence.

## Discussion

Elastofibroma is a benign, poorly circumscribed, tumor-like condition involving, in the vast majority of cases, the subscapular region of elderly individuals, though isolated cases have been seen in the deltoid muscle, infraolecranon area, hip, thigh and stomach (13). Elastofibromas are typically located in front of the scapula at a level corresponding to the sixth through to the eighth ribs, well under the serratus anterior, latissimus dorsi and levator scapulae muscles (13). Elastofibromas have been reported in other locations, and multiple elastofibromas have been described in the scapula and olecranon, and in the scapula and ischium (4), whereas reports of multiple elastofibromas in the same patient are rare.

Although the pathogenesis of elastofibromas is still unclear, repetitive microtrauma caused by friction between the scapula and the thoracic wall is known to cause reactive hyperproliferation of fibroelastic tissue (2, 14, 15). A systematic review of the literature did not shed any light on the role of microtrauma because most authors did not provide any information on their patients’ activities. A predominance of women over men ranging from 5:4 to as much as 13:1, depending on the study, suggests that microtrauma alone is unlikely to be the main factor underlying this lesion (3-6). Furthermore, more recent reports have reported a predominance in men. Giebel *et al.* (9) , in a series of 100 autopsies, found 13 patients with elastofibroma dorsi, 10 of whom were men (17%) and three women (7.3%), while Kransdorf *et al.* (16) found that men outnumbered women by 4:3. Previous studies have reported other sites exposed to friction, such as the tricuspid valve, axilla, foot and ischial tuberosity; reports of less common locations associated with lower mechanical stress, such as the mediastinum, the stomach, the greater omentum, the inguinal region, the orbita and the intraspinal space, also seem to suggest that elastofibromas are not caused by microtrauma (5, 14, 17). The tumors have been reported to occur in both the dominant and non-dominant side; if it is

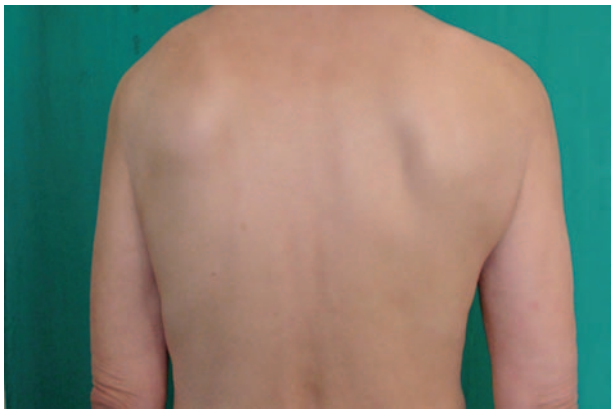

Figure 1. *Presence of a swelling measuring about 7×7 cm in the right subscapular region.*

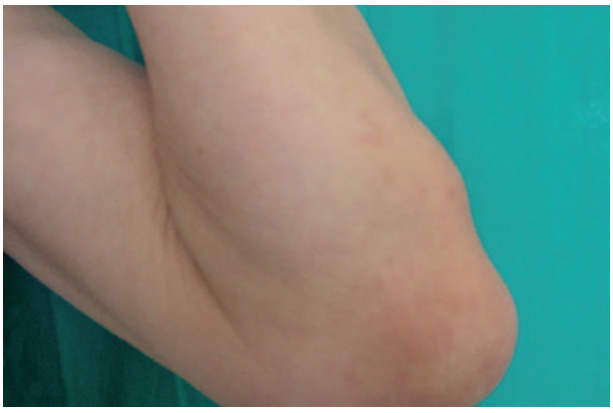

Figure 2. *Presence of a swelling measuring about 5×5 cm in the left elbow.*

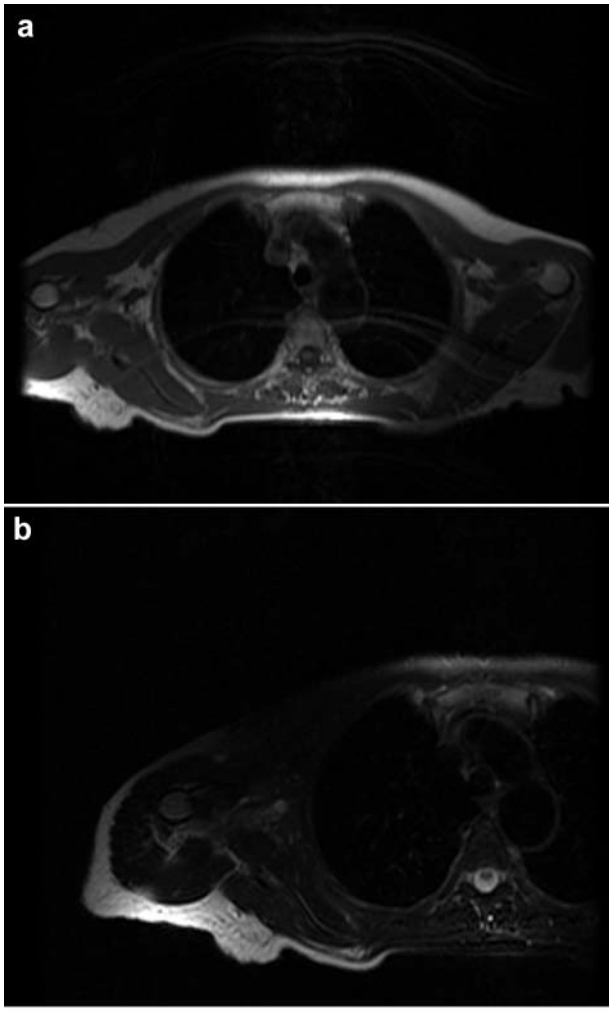


Figure 3. *CT imaging of the thorax using the multislice spiral technique: presence of solid soft tissue masses with well-defined margins and a heterogeneous signal intensity in the subscapular area.*

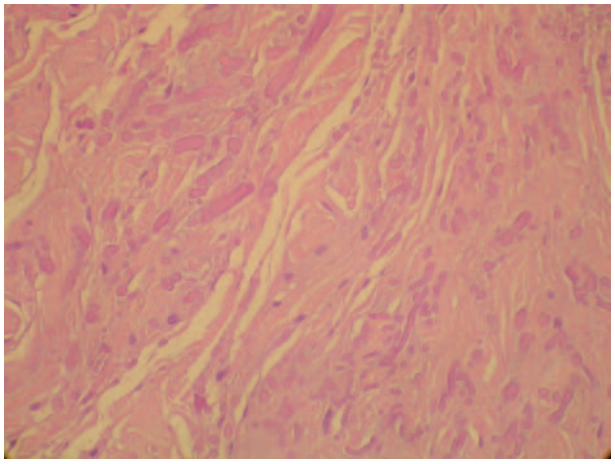

Figure 4. *Histopathological investigation: presence of fibrous, collagenous strands and plump, occasionally elongated, mostly round-shaped elastic fibres, which were densely packed. The elastic structures typically form discs or globules and sometimes appear in an "asbestos-body-like" fashion. The lesions are predominantly hypocellular with fibrocytic and fibroblastic cells without atypia or mitotic activity. (Haematoxylin and eosin, original magnification ×40).*

assumed that the dominant side was exposed to a higher level of repetitive microtrauma during the patient's lifetime, it may be concluded that there is no association between elstofibroma and mechanical stress. Our patient did not have a history of extensive physical activity either. Several authors have proposed vascular insufficiency as a possible reason for the degenerative changes (10, 15). A familial predisposition with an underlying enzymatic defect may exist in 30% of cases, though this has not yet been proven (4, 18). Large case series from Japan strongly suggest that hereditary factors may predispose to this lesion (4).

The nature of the altered elastic fibres has yet to be fully elucidated and remains a matter of debate. They may be caused by either abnormal elastogenesis or secondary degeneration, or by a combination of both (10, 12). The symptoms of elastofibroma dorsi depend on the site and size of the lesion and may present as shoulder and olecranon pain or snapping scapula, as occurred in this patient. In 50% of cases, the tumor remains asymptomatic or causes mild discomfort, which explains why there may be time lapses of up to 67 years between the onset of symptoms and treatment (3-6, 11). Large lesions may simulate scapula alata by raising the scapula. If palpable, the tumor may mimic semi-mobility owing to its elastic fibres, though it is usually found to be adherent to the surrounding tissue intra-operatively. While occurring predominantly on the right side, it may be bilateral in as many as 66% of cases (4, 10, 12). The concomitance of hypertension and dyspnea with elastofibroma has not previously been reported and may be highly unlikely, though a large tumor may impair chest elasticity and movements, thereby hampering breathing motor function and causing dyspnea.

If a possible soft tissue signal intensity or elevated scapula are excluded, plain radiographs do not show specific changes. On CT, the lesion signal intensity is usually comparable to that of muscle, margins are well defined and signal intensity tends to be low. Interspersed adipose strands cause a heterogeneous structure with longitudinal areas of higher signal intensity (6, 14, 19). In our patient, the CT findings were consistent with the aforementioned criteria. Faint, or even marked enhancement, mimicking malignancy may be observed after contrast agent administration (16, 20, 21). MRI, which is probably the most reliable non-invasive diagnostic technique, yields the same findings, but is more sensitive to the strands of fatty tissue (19). In this case, the diagnosis of elastofibroma dorsi was made on the basis of the typical subscapular location of the lesions and the characteristic CT appearance (poorly circumscribed soft tissue with attenuation similar to that of muscle). This CT finding was particularly relevant because the presumptive diagnosis of elastofibroma dorsi meant that a wide margin around the lesion was not required when surgery was performed. Radiotracer accumulation of the hypermetabolic tumor has been reported when PET-CT is used (22).

The fact that multicenter and familial cases have been described points to the existence of a predisposing background, with a history of intense manual labor often being reported. At surgery, the lesions are usually found at the apex of the scapula, beneath the rhomboid and latissimus dorsi muscles. The right side is more often affected than the left, and bilaterality is frequent. A periosteal origin has been suggested.

The differential diagnosis of elastofibromas includes sarcomas, aggressive fibromatosis, lipoma and fibroma. Owing to its muscle-like appearance in all the afore-mentioned imaging procedures, the lesion may go undiagnosed or may, in the presence of abnormal features, be misdiagnosed. The advanced age of the patients, the typical location, or bilateral manifestation should support a presumptive diagnosis of elastofibroma. One may refrain from performing a biopsy when all these features are present and the imaging findings are clear. An open biopsy, or at least a core needle biopsy, should be performed, if necessary, to obtain a representative tissue specimen. Some authors state that a diagnosis of elastofibroma dorsi should not be based on imaging features alone, a biopsy being required to rule out more aggressive tumors (23). According to other authors, no further tests are needed to make a diagnosis of elastofibroma dorsi when a mass of interposed fat and soft tissue is located deeply in relation to the scapula (24).

Microscopically, collagen bundles are alternated with numerous acidophilic, refractive cylinders that often contain a central dense core, both of which stain strongly with elastic stains. Ultrastructurally, the cylinders are made of immature amorphous elastic tissue, whereas the central core contains mature fibers. Elastase digestion fully removes this material. The biochemical composition of the collagen bundles is that of elastin, though its amino acid composition differs slightly from that of normal elastic tissue. The collagen deposited in the lesion is a mixture of types I, II and III; the presence of type II collagen is somewhat perplexing because this is normally only present in articular cartilage and some ocular structures. This lesion is not a true neoplasm but rather a reactive hyperplasia involving abnormal elastogenesis; the new material synthesized by the tumor cells appears to be laid down around pre-existing elastic fibers (13, 25).

When asymptomatic lesions are diagnosed incidentally, excision is not required as malignant transformation has never been reported. Only in cases of discomfort, scapula snapping or blocking and pain, is marginal resection strongly recommended, according to the patient's psychological and physical strain (3, 4, 26). Anecdotal reports have suggested that radiotherapy is also effective (17). Radiotherapy may be particularly indicated when the lesions are in unresectable locations.

In the presented case, both the subscapular and olecranon lesions were removed because the patient's symptoms could be ascribed to the areas involved. The patient's postoperative course was uneventful. The patient was disease-free at the follow-up; indeed, there are few reports of recurrences in the literature (4, 16).

It is recommended that elastofibroma be considered by both the surgeon and the surgical pathologist as a likely diagnosis in the differential diagnosis of soft tissue tumors, particularly those located in the subscapular region. CT and MRI can effectively be used to locate and identify the lesion. Unnecessary wide, radical resections should be avoided in symptomatic patients who undergo surgery because marginal resection has proved to be sufficient.

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