Pericytic / perivascular neoplasms have traditionally been dominated by haemangiopericytoma. However, it is now recognized that the latter diagnostic category subsumes a wide variety of tumour types which share the presence of thin-walled branching blood vessels. If such lesions are otherwise classified, there remains only a small group of spindle cell lesions designated as haemangiopericytoma, although they have no evident relationship to pericytes, and may be more closely related to solitary fibrous tumour (see Chapter 2).

The lesions now remaining in this pericytic / perivascular category all show evidence of differentiation towards myoid / contractile perivascular cells and all share the characteristic tendency to grow in a circumferential perivascular fashion. Currently, the term 'myopericytoma' is preferred to avoid confusion with the ill-defined former terminology.

Important advances have been made in predicting biological potential of glomus tumours and in understanding the close relationship between myopericytoma, myofibroma / myofibromatosis, and so-called infantile haemangiopericytoma, which essentially form a single morphological continuum. Their myoid nature and shared features with angioleiomyoma explain their more logical alignment with smooth muscle tumours rather than vascular tumours in this new classification.

Sinonasal haemangiopericytoma, which appears to be a truly pericytic lesion, is described in the Respiratory System volume.
Glomus tumours

Definition
Glomus tumours are mesenchymal neo-
plasms composed of cells that closely
resemble the modified smooth muscle
cells of the normal glomus body.

ICD-O codes
Glomus tumour 8711/0
Glomus tumours of uncertain
malignant potential 8711/1
Malignant glomus tumour 8711/3

Epidemiology
Glomus tumours are rare, accounting
for less than 2% of soft tissue tumours
[1946]. Multiple lesions may be seen in
close to 10% of patients. Malignant
glomus tumours are exceedingly rare,
comprising less than 1% of glomus tumours [697].
Glomus tumours typically occur in
young adults but may occur at any age.
No sex predilection is seen, except in
subungual lesions, which are far more
common in women (2079,2177).

Sites of involvement
The vast majority of glomus tumours
occur in the distal extremities, particular-
ly the subungual region, the hand, the
wrist and the foot (2246). Rare tumours
have however been reported in almost
every location, including the stomach (885),
penis (1132), mediastinum (952),
nerve (293), bone (1815) and lung (751).
Glomus tumours almost always occur in
the skin or superficial soft tissues,
although rare cases occur in deep soft
tissue or viscera. Malignant glomus
tumours are usually deeply seated, but
may be cutaneous (697).

Clinical features
Cutaneous glomus tumours are typically
small (<1 cm), red-blue nodules that are
often associated with a long history of
pain, particularly with exposure to cold
or minor tactile stimulation.
Deeply seated or visceral glomus
tumours may have either no associated
symptoms or symptoms referable to the
involved organ.

Histopathology

Typical glomus tumours
Typical glomus tumours are subcatego-
rized as “solid glomus tumour”, “glomang-
gioma”, and “glomangiomyoma” depend-
ning on the relative prominence of glomus
cells, vascular structures and smooth
muscle. Glomus cells are small, uniform,
rounded cells with a centrally placed,
round nucleus and amphophilic to lightly
eosinophilic cytoplasm. Each cell is sur-
rounded by basal lamina, seen best on
PAS or toluidine blue histochemical
stains. Occasionally cases show onco-
cytic (1967) or epithelioid change
(1737).

Solid glomus tumours are the most com-
mon variant, comprising approximately
75% of cases (2242). They are com-
posed of nests of glomus cells surround-
ing capillary sized vessels. The stroma
may show hyalinization or myxoid
change. Small cuffs of glomus cells are
often seen around small vessels located
outside of the main mass. Glomang-
giomas, comprising approximately 20%
of glomus tumours, are characterized by
dilated veins surrounded by small clus-
ters of glomus cells. Glomangiomas are
the most common type of glomus tumour
in patients with multiple or familial
lesions. Glomangiomyomas, the least
common subtype of typical glomus
tumour, are characterized by an overall
architecture similar to solid glomus
tumour or glomangioma and by a transi-
tion from typical glomus cells to elongat-
ed cells resembling mature smooth mus-
cle. In some glomus tumours a branch-
ing, haemangiopericytoma-like vascular-
ture is present and such cases have
been designated “glomangiopericytoma”
(825).

Symplastic glomus tumours
Symplastic glomus tumours show strik-
ing nuclear atypia in the absence of any
other worrisome feature (e.g., large size,
deep location, mitotic activity, necrosis)
(697). The marked nuclear atypia that
characterizes these tumours is believed
to be a degenerative phenomenon. All
cases reported to date have behaved in
a benign fashion.

Malignant glomus tumours
(glomangiosarcomas) and glomus
tumours of uncertain malignant potential
Histologically malignant glomus
tumours are exceedingly rare and clini-
cally malignant ones (e.g., metastatic)
rarer yet. Prior to 2000, fewer than 20
histologically malignant and 2 clinically
malignant tumours had been reported
(21,54,247,823,885,952,953,1575,
2219,2220, 2255). Criteria for the diag-
nosis of malignancy in glomus tumours
were only recently elaborated (697).
The diagnosis of “malignant glomus
tumour” should be reserved for tumours
showing: 1) Size >2 cm and subfascial
or visceral location; 2) Atypical mitotic
figures; or 3) Marked nuclear atypia and
any level of mitotic activity. These
features frequently co-vary in a given
case. A component of pre-existing
benign-appearing glomus tumour is
often but not always present. There are
two types of malignant glomus tumour.

Fig. 5.01 Glomus tumour. Note the typical rounded
cytomorphology and well defined cell membranes.

136 Pericytic (perivascular) tumours
In the first type, the malignant component resembles a leiomyosarcoma or fibrosarcoma. In the second type, the malignant component retains an overall architectural similarity to benign glomus tumour and consists of sheets of highly malignant appearing round cells. Immunohistochemical demonstration of smooth muscle actin and pericellular type IV collagen is required for the diagnosis of this second type of malignant glomus tumour, in the absence of a clear-cut benign precursor. Malignant glomus tumours are highly aggressive with metastases in approximately 40% of cases, resulting in the death of the patient [697]. Glomus tumours not fulfilling criteria for malignancy, but having at least one atypical feature other than nuclear pleomorphism should be diagnosed as “glomus tumours of uncertain malignant potential”.

**Immunohistochemistry**

Glomus tumours of all types typically express smooth muscle actin and have abundant pericellular type IV collagen production. H-caldesmon is also positive. Other markers, including desmin, CD34, cytokeratin and S100 protein are usually negative [697].

**Ultrastructure**

Ultrastructurally glomus cells have short interdigitating cytoplasmic processes, bundles of thin actin-like filaments with dense bodies and occasional attachments plaques to the cytoplasmic membrane and prominent external lamina [1449].

**Genetics**

Multiple familial glomus tumours appear to have an autosomal dominant pattern of inheritance [164,884,1363]. An association between subungual glomus tumours and neurofibromatosis type I has been reported [1109,1602,1867]. The gene for multiple inherited glomus tumours has been linked to chromosome 1p21-22 [229,297]. The genetic events underlying sporadic glomus tumours are not known.
Myopericytoma

Definition
Myopericytoma is a benign, generally subcutaneous tumour that is composed of oval-to-spindle shaped myoid appearing cells with a striking tendency for concentric perivascular growth. It is believed that the lesional cells show apparent differentiation towards perivascular myoid cells or myopericytes. Myopericytoma forms a morphological continuum with myofibroma, angioleiomyoma and so-called infantile haemangiopericytoma.

ICD-O code 8713/1

Synonyms
In the past, myopericytoma may have been diagnosed as a solitary myofibroma or “haemangiopericytoma.”

Epidemiology
Myopericytoma arises most commonly in mid adulthood; however, lesions can arise at any age. Familial cases have not been reported.

Sites of involvement
Myopericytoma generally arises in subcutaneous tissue. There is a predilection for lesions to involve the distal extremities; however, tumours can also arise at other sites, including the proximal extremities and neck. It is likely that a wider site distribution will be described with increased recognition of this tumour.

Clinical features
Myopericytoma generally presents as a painless, slow-growing subcutaneous nodule that can be present for years. Some lesions are painful. Myopericytoma most commonly arises as a solitary lesion but multiple lesions are not infrequent. Multiple lesions generally arise metachronously and usually involve a particular anatomic region such as a foot.

Macroscopy
Myopericytoma tends to be a well circumscribed nodule measuring less than 2 cm in diameter.

Histopathology
Myopericytomas are unencapsulated and most lesions are fairly well circumscribed. Lesions are composed of relatively monomorphic oval-to-spindle shaped myoid appearing cells that show striking multilayered concentric growth around lesional blood vessels. The cells have eosinophilic or amphophilic cytoplasm. Lesions can be solidly cellular; however some cases have prominent myxoid stroma. In occasional cases, the spindle cells fall apart in the intervascular regions. In many cases, blood vessels outside the
lesion also show concentric perivascular proliferation of spindle cells. Lesional blood vessels tend to be numerous and can be variable in size. In some cases, numerous thin walled branching or gaping blood vessels are present. Fascicular or whorled arrangements of spindle cells with abundant eosinophilic cytoplasm, embedded in myxoid stroma, are present in some cases. These areas are similar to the myoid whorls of myofibromatosis/myofibroma and invagination or bulging of these areas into the lumen of lesional blood vessels is frequently seen. Subendothelial proliferation of lesional cells in vessel walls is frequently seen and, indeed, myopericytoma can be located entirely within the lumen of a vein. Some myopericytomas have a component of cells with glomus-type features including cuboidal shape, distinct cell borders, clear to eosinophilic cytoplasm and central round nuclei and the term glomangiofibrocytoma can be used in such cases. In reality a spectrum of lesions exists that includes myofibromatosis, myofibroma, infantile haemangiopericytoma, glomangiofibrocytoma and myopericytoma (295.825). Rarely, lesions show marked hyalinization, cystic change or focal metaplastic bone. Mitoses are not conspicuous (generally much less than 1/10 HPF). Coagulative necrosis has been described in a glomangiofibrocytoma; however, this appears to be a very unusual finding (825).

**Immunophenotype**

The spindle cells in myopericytomas are positive for smooth muscle actin (SMA). SMA staining is generally diffusely positive, but can be only focally positive, generally in a perivascular distribution.

Occasional cases are focally desmin positive (825). Focal CD34 staining by lesional cells occurs in some cases. Lesional cells are negative for S100 protein and most cases are negative for cytokeratin.

**Prognostic factors**

Most myopericytomas do not recur following excision. Recurrence may be related to poor circumscription of a lesion. Sometimes it is difficult to know whether a myopericytoma has recurred or whether a new lesion has developed in the same anatomic area. Very rare malignant myopericytomas exist (1383).