7 Plasma Cell Proliferations

7.1 Plasma Cell Myeloma/Bone Marrow Plasmacytoma (ICD-O: 9732/3)

Synonyms
- KIEL: Plasmacytic lymphoma (plasmacytoma)
- REAL: Plasmacytoma /plasma cell myeloma
- WHO: Plasma cell myeloma
- Other: Multiple myeloma, Kahler’s disease

Definition
This neoplastic proliferation consists of a unique diagnostic triad of osteolytic bone lesions, marrow infiltration by atypical plasma cells, and serum monoclonal gammopathy (Kyle 1992; Schajowicz 1994; Malpas et al. 1995; Grogan et al. 2001).

Morphology

Macroscopy. The neoplasia is responsible for the development of nodules in the bone marrow that consist of soft grayish or yellowish homogeneous tissue and measure about 1 cm in diameter. These nodules transform into large polycyclic masses that destroy not only the bone architecture of the medulla but also the cortex with extension to the surrounding soft tissues. These bone lesions cause collapse of the vertebral bodies and fracture of tubular bones.

Histology. The diagnosis of plasma cell myeloma is based on the type of bone marrow infiltration, the quantity of neoplastic cells, and the morphology of the cells (Reed et al. 1981; Kyle 1992; Bartl et al. 1982, 1995; Sailer et al. 1995).

Early lesions are made up of small nests of neoplastic cells that are dispersed between adipose cells, more or less intermingled with hematopoietic cells, and at a distance from the arterioles (Fig. 7.1). This localized interstitial involvement can be difficult to diagnose. The presence of large plasma cells, blastic or pleomorphic variants, is useful in making the diagnosis.

The presence of aggregate of about 25 plasma cells (Grogan et al. 2001) as well as of more diffuse interstitial infiltrates (Fig. 7.2) favors the diagnosis of plasma cell myeloma. Some of the aggregate may develop along the bone trabeculae (Fig. 7.3).

In more advanced cases, sheets of plasma cells form nodules or strands destroying and replacing the hematopoietic tissue. Finally, densely packed plasma cells lead to massive involvement of the medullary spaces. Collagen bands can develop around vessels and bone trabeculae (Fig. 7.3). Destruction of bone trabeculae by osteoclastic hyperplasia is often observed, mostly in advanced disease.

The neoplastic cells exhibit a variety of morphologies that either predominate or are variably associated. One of the most frequent neoplastic cell types is that of mature plasma cells (Fig. 7.1), Marschalko type; these cells have an eccentric nucleus with dense chromatin (“cartwheel” pattern), no or only one small nucleolus, a large ovoid or triangular basophilic cytoplasm, and a clear juxtanuclear halo (extremely large Golgi apparatus). Giant forms can be observed.

Another neoplastic cell type is lymphoid plasma cells. These are smaller than Marschalko type cells, with scanty cytoplasm surrounding a round nucleus with dense chromatin blocks. They seem to often be associated with IgD production (Reed et al. 1981).

Proplasma cells are larger, with a pale nucleus containing a central medium-sized nucleolus (Fig. 7.4). Other types of plasma cells have an irregular, “cleaved” or “notched” nucleus. Large cells with the morphology of immunoblasts, plasmablasts (Fig. 7.5), or even giant multinucleated cells mimicking Sternberg-Reed cells are also observed. Vacuoles may be located in the cyto-
Fig. 7.1. Plasma cell myeloma. Early infiltration by dispersed small nests of mature plasma cells (Marschalko type), at a distance from capillaries and arterioles (Giemsa stain).

Fig. 7.2. Plasma cell myeloma. Interstitial infiltrate by plasma cells replaces the normal hematopoietic cells (H and E stain).