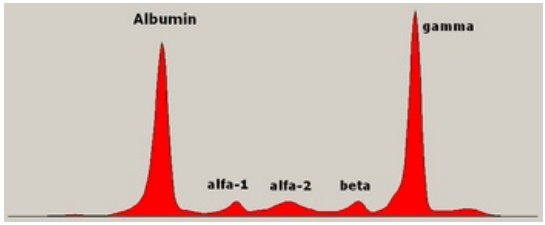
Paraproteinemia

Increased concentration of monoclonal immunoglobulins or their fragments in serum. These immunoglobulins are called **paraproteins**', or the so-called **M-component**' (M - monoclonal). Monoclonality of proteins is caused by excessive multiplication of a single clone of plasmocytes, often on the basis of malignant growth - plasmocytomau. Depending on the cell type, complete immunoglobulins of the class IgG or IgM, or only light or heavy chains are synthesized.



Electrophoresis of proteins in multiple myeloma

The presence of paraproteins causes the formation of a narrow and high peak in the area of γ-globulins during serum protein electrophoresis.

If only light chains of immunoglobulins are synthesized, these penetrate the glomerular membrane into the urine, where they can be detected as the so-called *Bence-Jones protein*. This protein is not detectable with ordinary test strips - electrophoresis of urine or its heating (protein denaturation) is required for capture. Bence-Jones protein can cause kidney damage. Cysts may form in the distal tubules and cause nephropathy, or the formation of crystals in the cytoplasm of proximal tubule cells may cause Fanconi syndrome.

The concentration of paraprotein in the blood reflects the extent and activity of the pathological clone of cells. At a significant concentration, it causes the so-called **hyperviscosity syndrome** (impairment of vision, thrombosis, neurological symptoms). If the M-component has the character of cryoglobulins, specific microcirculation disorders arise, Raynaud's syndrome (cryoglobulins precipitate here in the cold acral parts of the body). At the same time, the formation of paraprotein is accompanied by a decrease in the formation of normal Ig and their increased degradation. Typically, resistance to infections is reduced.

Paraprotein formation is characteristic of the following **lymphoproliferative syndromes**': multiple myeloma, Waldenström macroglobulinemia, primary amyloidosis, heavy chain disease, and MGUS (monoclonal gammopathy of uncertain severity).

Multiple Myeloma (Plasmocytoma)

Progressive damage to the skeleton by neoplastic proliferation of plasma cells, diffuse form. Increased production of IgG or IgA. Possible capture of Bence-Jones protein during the production of light chains in the urine. The liver, spleen, and lymph nodes are not enlarged. Bone tissue is increasingly resorbed by osteoclasts, rapid development of osteoporosis, destructive and osteolytic changes in axial parts of the skeleton, pathological fractures. Hypercalcemia due to paracrine production of osteolytic factors by malignant cells. Damage to renal tubular function (glycosuria, aminoaciduria, ...), secondary anemia normocytic and normochromic.

Waldenström macroglobulinemia

Neoplastic proliferation of plasmacytes producing IgM. Hepatomegaly, splenomegaly, and lymphadenopathy are often present. Bone metabolism is not disturbed, there is no proteinuria. Present cryoglobulin, Raynaud's syndrome, anemia, hemorrhagic diathesis.

Primary amyloidosis

It actually arises on the basis of multiple myeloma. Amyloid fibrils contain parts of Ig light chains or whole light chains or parts of heavy chains. The source is a pathological clone of plasma cells. Non-specific symptoms (fatigue, weight loss, syncopes...).

Heavy chain disease

Pathological production of parts of Ig heavy chains that are present in plasma and urine. There are three forms of the disease (according to the types of heavy chains): γ, α, μ. It may accompany lymphoma or other lymphoproliferative diseases.

MGUS - monoclonal gammopathy of uncertain severity

Expansion of a single clone of plasma cells that, for unknown reasons, does not behave malignantly. It most commonly involves the production of IgG and IgA. Fairly common, requires monitoring.

Links

Related links

- Dysproteinemia
- Plasma proteins
- Hypergammaglobulinemia