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Mucinous Adenocarcinoma on Perianal Fistula

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Clinical Note

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Mucinous adenocarcinoma on perianl fistula is extremely rare entity (2- 3%) of all digestive malignancies). But we have recently diagnosed several cases in a few months. We have questioned ourselves whether it is a disease that is raising its frequency or simply it has been miss diagnosed in the past. New generation magnetic resonances are able to accurate make the statement and localize exactly where the origin of the tumour is. It may be due to either dysplastic degeneration in a long-standing recurrent fistula or seeding of granulating fistula by malignant cells arising from carcinoma originating from colorectal mucosa. We notice the need of point that this disease is usually of worse prognostic than the regular adenocarcinoma of the anal canal. Two questions can arise from this fact:

- 1. Is it really a different disease?
- 2. Is it usually diagnosed later than others?

It is known that sometimes endoscopy is normal because they have its origin in a perianal fistula. Most patients have undergone several perianal surgeries. Anal fistula reparation, drainage of perianal abscess, etc... And it is often retarded the real diagnose. Often biopsies are not taken because not malignancy is suspected. Chronic inflammatory tissue is a known premalignant factor as it is in esophageal cancer and other malignancies. Pilonidal disease is similar in this sense and also is perianal Crohn's disease. It is advisable to take samples for biopsies when these patients are operated on. We have to suspect a malignancy especially when the fistula is recurrent.

The prognosis of perianal cancer depends on the stage at diagnosis. MRI is the main imaging method for staging. Endoanal ultrasound can help in local staging, but since the mucosa is not affected, it is not as determinant as in rectal cancer.

The preoperative staging should be established as in anal cancer. Dimension of the tumor is the main factor in the prognosis. These tumours should be considered very orally aggressive tumours. So preoperative neoadjuvant therapy is often needed. Also surgery must be aggressive because sphincter muscle is often involved and must be removed. Abdominoperineal resection is the standard operation for these cases. The lymphatic dissemination can be as much towards the mesenteric ganglia as towards the ganglionic iliac chain. Some authors recommend a systematic PET scan to look for uptake in the iliac chain. Other authors advocate systematic iliac chain lymphadenectomy. Pathology specimen usually reveals dilated tortous glands with lakes of mucin and intervening stroma showing moderate lymphoplasmacitic infiltration and desmoplastic reaction consistent with mocinous adenocarcinoma.

Survival in this type of tumors is similar to rectal cancer. Although local recurrence seems to be more frequent.

References

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