

Portsmouth Regional Hospital
Cancer Program
Experience with Gastrointestinal Stromal Tumors
Between the Years 2004 and 2009

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REASON FOR REPORT: The reason for this report is to identify what appears to be some increased number of diagnosed GIST tumors found at the Portsmouth Regional Hospital Cancer Program. On review of the literature, it seems that we possibly have a higher rate than we did in the past and nationally, although it is hard to know for sure.

HISTORY OF GIST TUMORS: A GIST tumor is a soft tissue sarcoma recently diagnosed as being separate from a leiomyosarcoma for some of the testing. Soft tissue sarcomas in general represent about 1% of neoplasms in the adult. The GIST account for 25% of the sarcomas. The GIST tumor is a neoplasm of the intestinal cell coming from the Cajal intestinal cell. This cell plays a role in intestinal motility. The GIST tumor is the most common solid tumor found in the GI tract. Until recently, surgery was the only means of cure although there have been some recent changes for chemotherapy intervention.

Most GIST tumors have a gain of function mutation found in the C-kit protogenetic site which then stimulates anopposed cell growth. The GIST tumor was historically unknown in the past because GIST tumors were considered benign or leiomyomas or leiomyosarcomas. However, recently GIST tumors have become more commonly diagnosed because of the of the C-kit testing. It is felt in some of the studies to be at least 1,000 cases of GIST diagnoses per year although probably larger as we become more familiar with this tumor. The GIST tumor most commonly arises from the stomach, followed by the small bowel, then colon and rectum, although the colon and rectum or esophageal tumors are fairly rare. However, even rarer, they can occur in the retroperitoneum.

The age distribution tends to be between 40 and 80 years the median being 58 years. It seems it is more prevalent in males than females. The presentation tends to be silent

although they can have symptoms in larger tumors with nonspecific abdominal complaints with abdominal cramps although 25% do present as bleeding.

The therapy for GIST tumors is surgical resection. The recommendation for surgical resection includes avoiding tumor rupture during the removal of the tumor. Since it rarely metastasizes, lymph node resection is not necessary. Local resection tends to be reasonable if not diffusely infiltrating into other tissues which is usually uncommon.

The prognosis of GIST tumors depends on the size and mitotic findings. If the tumor size is greater than 10 cm, it tends to have a 20% five-year survival rate. The survival rate goes down if the tumor ruptures. The prognosis is worse if there is a high mitotic rate, such as aneuploidy cellular morphology and proliferative index and assays.

Recent development of the STI-571 (imatinib), otherwise known as Gleevec, has changed the therapy for GIST tumors. The Gleevec inhibits the site of platelet-derived growth factor area. Presently, Gleevec has been used in metastatic tumors found at presentation. It is becoming more and more prevalent with C-kit positive GIST tumors. The therapy has become multidisciplinary with multidisciplinary specialists including medical oncologists, surgeons, and pathologists. The C-kit mutation is still unknown as the percentage that is present in the presenting tumor. The C-kit positive tumors tend to be more malignant than the C-kit negative tumors. Eighty-five percent of GIST tumors are C-kit positive. A recent Japanese study showed that a five-year survival rate with C-kit negative tumors is 86% and C-kit positive tumors 49% when present.

Of the experience at Portsmouth Regional Hospital analyzed between 2004 and 2009, thirteen patients were identified. Index 1 shows the number of patients, their age distribution, their sex distribution, location of the tumor found at diagnosis, the size of the tumor, its mitotic rate, its C-kit positive versus negative rate, and therefore its risk including some of the studies that have shown whether the tumor is vimentin positive versus negative and CD-35 findings. In the study, it also shows when the patient received Gleevec or not.

In the analysis, the age distribution from our study shows the age of 69 which is a little older than the national rate of 58. In our study, there is still a prevalence of male verses female. Only half the patients in our study were Gleevec positive. Because of the study being done between 2004 and 2009, we do not know yet our five-year survival rate. So far, there was only one death within the study, but this was not due to the tumor itself but to an underlying medical problem.

We will continue to watch the five-year survival rate on all patients studied so far. With the increasing awareness of GIST tumors and the increasing awareness of Gleevec therapy, we will continue to watch our outcomes accordingly.

CONCLUSION: We find a rise in GIST tumors at Portsmouth Regional Hospital. The enclosed study shows our distribution of the tumor. We find that the increased rate is secondary to the awareness of the tumors, especially with the C-kit positive status, and this is consistent with the trends nationally. We will continue to monitor the findings and the diagnosis and potential therapies in the future.