GISTs," ascribed to us by Manley and colleagues, is inaccurate. Actually, *PDGFRA*-mutant GISTs are in most cases not polypoid, rather constituting intramural masses, with clear-cut histologic and immunophenotypical features [3]. We have recently produced morphogenetic evidence of the origin of these tumors, along with typical IFPs, from TCs, exploiting the opportunity offered by the concurrent presence of a prominent TC hyperplasia [4]. In the same article, we proposed to define IFP as "telocytoma" because this term conveys both the neoplastic and histotypic natures of these tumors.

Conversely, tumors typical of *PDGFRA*-mutant syndrome whose nosology is less straightforward are the fibrous tumors defined as GISTs by De Raedt and colleagues [5] and described also by Carney and Stratakis [6] and by ourselves [7]. These tumors not only do not exhibit morphologic features typical of GISTs but also do not express CD117 and DOG1; in addition, they have been found mostly in the small intestine, unlike *PDGFRA*-mutant GISTs, which show a strong predilection for the stomach. We have previously discussed in depth this issue, supporting these "fibrous tumors" as a possible variant of IFP [7].

In conclusion, in our opinion, *PDGFRA* mutations can determine 3 types of lesions in the gastrointestinal tract: (1) typical *PDGFRA*-mutant GIST, (2) typical IFP (telocytoma) and its variant formerly defined as "fibrous tumors", and (3) TC hyperplasia. Of note, the latter and the "fibrous tumors" variant of IFP (telocytoma) hitherto have been described exclusively in germline *PDGFRA* mutant settings, that is, in *PDGFRA*-mutant syndrome.

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Telocytes as possible precursors of *PDGFRA*-mutant gastrointestinal mesenchymal tumors—reply to rejoinder

To the Editor,

We are flattered that Giustiniani et al have chosen to use the term "telocytoma," first mentioned in the literature by us in our reply to their letter [1], to describe inflammatory fibroid polyps (IFPs). This may prove a confusing term if *PDGFRA*-mutant gastrointestinal stromal tumors (GIST) also arise from the telocyte as they suggest.

If the cell of origin of gastric *PDGFRA*-mutant GISTs is the telocyte, the molecular pathogenesis is clearly more complex than that of the IFP. There are as yet no described gastric CD117- or DOG1-positive progenitor telocyte cells associated with these GISTs, suggesting that the cell of origin is not the mature telocyte, or that there are additional unknown sporadic mutations within the telocyte. We await the upcoming publication of Ricci et al [2]. We would also encourage others to study the skin in familial *PDGFRA* mutation patients with coarse facies and/or broad hands and evaluate the role of the telocyte in producing this particular phenotype.

In patients with a familial *PDGFRA* mutation, there is widespread PDGFRA overexpression resulting in variable hyperplasia of CD34, PDGFRA-positive telocyte cells in the stomach and intestine. The hyperplasia may take the form of firm flat masses in the intestine, called fibrous tumors by some, but these areas are often contiguous with elevated plateaus and/or polyps [3]. Because these shapes all directly reflect the PDGFRA-driven hyperplasia of telocytes, it is unnecessary to distinguish IFPs from fibrous tumors as Giustiniani and colleagues repeatedly do, although in their article, none of their IFPs have discrete borders [4].

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