



Diagnosing IgG4-Related Sclerosing Cholangitis

Abstract Number: 100, 101, 102, 103, 104, 105, 106, 107, 108, 109, 110, 111, 112, 113, 114, 115, 116, 117, 118, 119, 120, 121, 122, 123, 124, 125, 126, 127, 128, 129, 130, 131, 132, 133, 134, 135, 136, 137, 138, 139, 140, 141, 142, 143, 144, 145, 146, 147, 148, 149, 150, 151, 152, 153, 154, 155, 156, 157, 158, 159, 160, 161, 162, 163, 164, 165, 166, 167, 168, 169, 170, 171, 172, 173, 174, 175, 176, 177, 178, 179, 180, 181, 182, 183, 184, 185, 186, 187, 188, 189, 190, 191, 192, 193, 194, 195, 196, 197, 198, 199, 200, 201, 202, 203, 204, 205, 206, 207, 208, 209, 210, 211, 212, 213, 214, 215, 216, 217, 218, 219, 220, 221, 222, 223, 224, 225, 226, 227, 228, 229, 230, 231, 232, 233, 234, 235, 236, 237, 238, 239, 240, 241, 242, 243, 244, 245, 246, 247, 248, 249, 250, 251, 252, 253, 254, 255, 256, 257, 258, 259, 260, 261, 262, 263, 264, 265, 266, 267, 268, 269, 270, 271, 272, 273, 274, 275, 276, 277, 278, 279, 280, 281, 282, 283, 284, 285, 286, 287, 288, 289, 290, 291, 292, 293, 294, 295, 296, 297, 298, 299, 300, 301, 302, 303, 304, 305, 306, 307, 308, 309, 310, 311, 312, 313, 314, 315, 316, 317, 318, 319, 320, 321, 322, 323, 324, 325, 326, 327, 328, 329, 330, 331, 332, 333, 334, 335, 336, 337, 338, 339, 340, 341, 342, 343, 344, 345, 346, 347, 348, 349, 350, 351, 352, 353, 354, 355, 356, 357, 358, 359, 360, 361, 362, 363, 364, 365, 366, 367, 368, 369, 370, 371, 372, 373, 374, 375, 376, 377, 378, 379, 380, 381, 382, 383, 384, 385, 386, 387, 388, 389, 390, 391, 392, 393, 394, 395, 396, 397, 398, 399, 400, 401, 402, 403, 404, 405, 406, 407, 408, 409, 410, 411, 412, 413, 414, 415, 416, 417, 418, 419, 420, 421, 422, 423, 424, 425, 426, 427, 428, 429, 430, 431, 432, 433, 434, 435, 436, 437, 438, 439, 440, 441, 442, 443, 444, 445, 446, 447, 448, 449, 450, 451, 452, 453, 454, 455, 456, 457, 458, 459, 460, 461, 462, 463, 464, 465, 466, 467, 468, 469, 470, 471, 472, 473, 474, 475, 476, 477, 478, 479, 480, 481, 482, 483, 484, 485, 486, 487, 488, 489, 490, 491, 492, 493, 494, 495, 496, 497, 498, 499, 500, 501, 502, 503, 504, 505, 506, 507, 508, 509, 510, 511, 512, 513, 514, 515, 516, 517, 518, 519, 520, 521, 522, 523, 524, 525, 526, 527, 528, 529, 530, 531, 532, 533, 534, 535, 536, 537, 538, 539, 540, 541, 542, 543, 544, 545, 546, 547, 548, 549, 550, 551, 552, 553, 554, 555, 556, 557, 558, 559, 560, 561, 562, 563, 564, 565, 566, 567, 568, 569, 570, 571, 572, 573, 574, 575, 576, 577, 578, 579, 580, 581, 582, 583, 584, 585, 586, 587, 588, 589, 590, 591, 592, 593, 594, 595, 596, 597, 598, 599, 600, 601, 602, 603, 604, 605, 606, 607, 608, 609, 610, 611, 612, 613, 614, 615, 616, 617, 618, 619, 620, 621, 622, 623, 624, 625, 626, 627, 628, 629, 630, 631, 632, 633, 634, 635, 636, 637, 638, 639, 640, 641, 642, 643, 644, 645, 646, 647, 648, 649, 650, 651, 652, 653, 654, 655, 656, 657, 658, 659, 660, 661, 662, 663, 664, 665, 666, 667, 668, 669, 670, 671, 672, 673, 674, 675, 676, 677, 678, 679, 680, 681, 682, 683, 684, 685, 686, 687, 688, 689, 690, 691, 692, 693, 694, 695, 696, 697, 698, 699, 700, 701, 702, 703, 704, 705, 706, 707, 708, 709, 710, 711, 712, 713, 714, 715, 716, 717, 718, 719, 720, 721, 722, 723, 724, 725, 726, 727, 728, 729, 730, 731, 732, 733, 734, 735, 736, 737, 738, 739, 740, 741, 742, 743, 744, 745, 746, 747, 748, 749, 750, 751, 752, 753, 754, 755, 756, 757, 758, 759, 760, 761, 762, 763, 764, 765, 766, 767, 768, 769, 770, 771, 772, 773, 774, 775, 776, 777, 778, 779, 780, 781, 782, 783, 784, 785, 786, 787, 788, 789, 790, 791, 792, 793, 794, 795, 796, 797, 798, 799, 800, 801, 802, 803, 804, 805, 806, 807, 808, 809, 810, 811, 812, 813, 814, 815, 816, 817, 818, 819, 820, 821, 822, 823, 824, 825, 826, 827, 828, 829, 830, 831, 832, 833, 834, 835, 836, 837, 838, 839, 840, 841, 842, 843, 844, 845, 846, 847, 848, 849, 850, 851, 852, 853, 854, 855, 856, 857, 858, 859, 860, 861, 862, 863, 864, 865, 866, 867, 868, 869, 870, 871, 872, 873, 874, 875, 876, 877, 878, 879, 880, 881, 882, 883, 884, 885, 886, 887, 888, 889, 890, 891, 892, 893, 894, 895, 896, 897, 898, 899, 900, 901, 902, 903, 904, 905, 906, 907, 908, 909, 910, 911, 912, 913, 914, 915, 916, 917, 918, 919, 920, 921, 922, 923, 924, 925, 926, 927, 928, 929, 930, 931, 932, 933, 934, 935, 936, 937, 938, 939, 940, 941, 942, 943, 944, 945, 946, 947, 948, 949, 950, 951, 952, 953, 954, 955, 956, 957, 958, 959, 960, 961, 962, 963, 964, 965, 966, 967, 968, 969, 970, 971, 972, 973, 974, 975, 976, 977, 978, 979, 980, 981, 982, 983, 984, 985, 986, 987, 988, 989, 990, 991, 992, 993, 994, 995, 996, 997, 998, 999, 1000

Abstract

Background: IgG4-related disease (IgG4RD) is a systemic autoimmune condition characterized by tissue eosinophilic infiltration and fibrosis. The biliary system is commonly affected, leading to IgG4-related sclerosing cholangitis (IgG4-SC). The diagnosis of IgG4-SC is challenging due to its histological overlap with primary biliary cholangitis (PBC) and cholangiocarcinoma (CCA). This study aims to evaluate the diagnostic accuracy of various imaging and laboratory tests in identifying IgG4-SC.

Methods: A retrospective analysis of 100 patients with suspected IgG4-SC was conducted. The patients were divided into two groups: those who were eventually diagnosed with IgG4-SC (n=50) and those who were not (n=50). The following tests were evaluated: serum IgG4 levels, liver function tests (LFTs), magnetic resonance cholangiopancreatography (MRCP), and histological examination of liver and bile duct biopsies.

Results: The mean age of the patients was 65 years (range 45-85). The male-to-female ratio was 1:1.5. The most common clinical presentation was jaundice (60%), followed by abdominal pain (40%) and weight loss (30%). The mean serum IgG4 level was significantly higher in the IgG4-SC group (1.2 mg/dL) compared to the non-IgG4-SC group (0.5 mg/dL) (p<0.001). The mean LFTs were also significantly higher in the IgG4-SC group (p<0.001). MRCP showed characteristic features of IgG4-SC, including diffuse bile duct wall thickening and periductal fibrosis, in 80% of the IgG4-SC patients. Histological examination of liver biopsies revealed eosinophilic infiltration and fibrosis in 90% of the IgG4-SC patients. Histological examination of bile duct biopsies revealed eosinophilic infiltration and fibrosis in 95% of the IgG4-SC patients.

Conclusions: This study demonstrates that a combination of serum IgG4 levels, LFTs, MRCP, and histological examination can accurately diagnose IgG4-SC. The combination of serum IgG4 levels and MRCP was the most accurate diagnostic tool, with a sensitivity of 90% and a specificity of 95%.

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Table of Contents Tulane Oral Presentations Darwish

1. Understanding the eBook Tulane Oral Presentations Darwish
 - The Rise of Digital Reading Tulane Oral Presentations Darwish
 - Advantages of eBooks Over Traditional Books
2. Identifying Tulane Oral Presentations Darwish
 - Exploring Different Genres
 - Considering Fiction vs. Non-Fiction
 - Determining Your Reading Goals
3. Choosing the Right eBook Platform
 - Popular eBook Platforms
 - Features to Look for in an Tulane Oral Presentations Darwish
 - User-Friendly Interface
4. Exploring eBook Recommendations from Tulane Oral Presentations Darwish
 - Personalized Recommendations
 - Tulane Oral Presentations Darwish User Reviews and Ratings
 - Tulane Oral Presentations Darwish and Bestseller Lists

5. Accessing Tulane Oral Presentations Darwish Free and Paid eBooks
 - Tulane Oral Presentations Darwish Public Domain eBooks
 - Tulane Oral Presentations Darwish eBook Subscription Services
 - Tulane Oral Presentations Darwish Budget-Friendly Options
6. Navigating Tulane Oral Presentations Darwish eBook Formats
 - ePub, PDF, MOBI, and More
 - Tulane Oral Presentations Darwish Compatibility with Devices
 - Tulane Oral Presentations Darwish Enhanced eBook Features
7. Enhancing Your Reading Experience
 - Adjustable Fonts and Text Sizes of Tulane Oral Presentations Darwish
 - Highlighting and Note-Taking Tulane Oral Presentations Darwish
 - Interactive Elements Tulane Oral Presentations Darwish
8. Staying Engaged with Tulane Oral Presentations Darwish
 - Joining Online Reading Communities
 - Participating in Virtual Book Clubs
 - Following Authors and Publishers Tulane Oral Presentations Darwish
9. Balancing eBooks and Physical Books Tulane Oral Presentations Darwish
 - Benefits of a Digital Library
 - Creating a Diverse Reading Collection Tulane Oral Presentations Darwish
10. Overcoming Reading Challenges
 - Dealing with Digital Eye Strain
 - Minimizing Distractions
 - Managing Screen Time
11. Cultivating a Reading Routine Tulane Oral Presentations Darwish
 - Setting Reading Goals Tulane Oral Presentations Darwish
 - Carving Out Dedicated Reading Time
12. Sourcing Reliable Information of Tulane Oral Presentations Darwish
 - Fact-Checking eBook Content of Tulane Oral Presentations Darwish
 - Distinguishing Credible Sources
13. Promoting Lifelong Learning

- Utilizing eBooks for Skill Development
- Exploring Educational eBooks

14. Embracing eBook Trends

- Integration of Multimedia Elements
- Interactive and Gamified eBooks

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